Clinical Orthopaedics

ANTHONY F. DePALMA
Editor-in-Chief

With the Assistance of the
ASSOCIATE EDITORS
THE BOARD OF ADVISORY EDITORS
THE BOARD OF CORRESPONDING EDITORS



Number Eight
Fall, 1956



J. B. LIPPINCOTT COMPANY
Philadelphia and Montreal

Distributed in Great Britain by Pitman Medical Publishing Co., Limited, London

Library of Congress Catalog Card Number 53-7647

Clinical Orthopaedics is designed for the publication of original articles offering significant contributions to the advancement of surgical knowledge.

Original, typed manuscripts, not carbon copies, and illustrations should be forwarded prepaid to Dr. Anthony F. DePalma, 1025 Walnut Street, Philadelphia 7, Pa.

Manuscripts should be typed double spaced on one side of standard typewriter paper, leaving wide margins. While every effort will be made to guard against loss, it is advised that authors retain copies of manuscripts submitted. All pages should be numbered. Dorland's *American Illustrated Medical Dictionary* (edition 22) and Webster's *New International Dictionary* (edition 2) should be used as standard references. Scientific names for drugs should be used when possible. Copyright or trade names of drugs should be capitalized. Units of measurement, e.g., dosage, should be expressed in the metric system Temperature should be expressed in degrees centigrade. A contribution in a foreign language, when accepted, will be translated and published in English.

Black-and-white illustrations will be reproduced free of charge, but the publisher reserves the right to establish a reasonable limit upon the number. Ordinarily, colored illustrations cannot be published except at the author's expense. Black-and-white photographs should be in the form of glossy prints. *These should not be defaced in any way.* Any changes desired in them should be marked on a tissue overlay. This should be done before it is pasted to the print, since it is important not to mar the print in any way. Or any changes may be indicated on a separate sheet of paper. Line and wash drawings should be on white art board, with lettering in black India ink large enough to be readable after necessary reduction. Large or bulky illustrations should be accompanied by smaller glossy reproductions of the same to facilitate their circulation among the members of the editorial board. Illustrations should be numbered, the tops indicated, and the author's name and the title of the article in brief should appear on the back. *However, this should be done lightly, so as to leave no imprint on the face of the illustration.* A separate typewritten sheet of legends for the illustrations should be supplied.

A bibliography of numbered references *in alphabetic order* should appear at the end of the manuscript with corresponding numbering in the text. Bibliographies should conform to the style of the *Quarterly Cumulative Index Medicus.*

If a book:

Author's name, title of book, edition if there is [illegible] order named.

If an article in a journal:

Author's name, title of article, volume number, inclusive page numbers, year of publication, in the order named.

Manuscript may be submitted to us in the original language of the author. Now it is our policy to [illegible] lingua.

Following are the general subjects of forthcoming issues of *Clinical Orthopaedics:*

The Pathologic Physiology of Metabolic Bone Diseases, Spring, 1957
Affections of Growth Centers (Epiphyses, Apophyses), Fall, 1957
Orthopaedic Surgery in the Geriatric Patient, Spring, 1958
Rehabilitation, Fall, 1958

Contents

Section I

CHRONIC HEREDITARY DISEASES AND DEVELOPMENTAL ANOMALIES

Section II

GENERAL ORTHOPAEDICS

Section III

MOTORIST INJURIES AND MOTORIST SAFETY

Reduction of Injuries (Crash-Impact Engineering)

(Part 2)

Guest Editor: Jacob Kulowski, M.D.
Saint Joseph, Missouri

SECTION I

CHRONIC HEREDITARY DISEASES AND DEVELOPMENT ANOMALIES

Foreword

The uniqueness of man must be kept constantly in mind when investigating correlations and causes of *hereditary diseases and congenital malformations*. Every year more complex factors are related to human mutation rates and methods of detecting genetic carriers in man. Genetists are inclined toward a reduction in estimates of mutation rates in some disorders; for example, in the chondrodystrophies, recently computed as 1 in 100,000, and the cerebral palsies, as 1 in 1,000, of the present population. On the other hand, the mutation rate for fibrosis of the pancreas is computed as 1 in 300, a very high rate.

Most hereditary investigations are directed toward the more easily observed disorders affecting the integumentary and the endoskeletal systems. Accordingly, orthopaedics has profited as much as any other medical area from such research. The list of musculoskeletal disorders reported in the genetic literature is increasingly large. Hypotheses relate these disorders to basic metabolic disturbances of many varieties.

Morphogenetic studies of the rabbit reported recently by Crary and Sawin (1955), of the Roscoe B. Jackson Memorial Laboratory, Bar Harbor, indicate a racial difference in regional growth at the onset of vertebral ossification. These localized areas of greater or of lesser growth supply clues to the origin of adult skeletal and vascular variations. In hybrids between these races of rabbits, as observed in the newborn and in adults, such areas of growth differentiation blend or interact, and in subsequent generations there is evidence of genetic segregation. The findings of Sawin and colleagues suggest that body variations of bone and blood vessels may occur with greater frequency in some races (related groups of individuals in a breeding population), as do also definite patterns of retarded or accelerated regional growth of vertebral structures. These may appear as additional or supernumerary ribs, articulations, vertebrae or sternebrae and, by deduction, of abnormal muscle patterns and nerves. When growth is altered asymmetrically, they may appear as hemivertebrae, wedged vertebrae or divided vertebrae. Growth is considered to be retarded in the embryo rabbit when ossification is initiated. Furthermore, any growth retardation or acceleration within the developmental pattern, activated by genetic or environmental agents, will be greatest in those regions or areas of growth most active at the time of application. Of course this applies only to prenatal, not to postnatal, growth. Many varieties of body form, structural differences and anomalies may follow.

Specifically, such hereditary studies may be of great significance in determining some of the basic correlations of scoliosis. If there are races of rabbits that show a variety of scoliotic patterns, probably of a genetic character, they may open approaches that will throw light on these disorders in man. Instead of consuming energy in pursuing useless and untimely eugenic pathways, greater profit should come from basic research along lines of present-day nutritional, environmental and morphologic investigations. With a marked drop in infant mortality during recent years, accompanied by a child population under 16 years of age that has increased to 54 millions, the extent of our problem is apparent. We must explore every approach with the aid of fellow scientists in other disciplines.

CHARLES W. GOFF

1

Louis Bauer, Orthopaedist Extraordinary

CHARLES W. GOFF, M.D.*

In 1869 St. Louis was a wild town. Texas punchers roared in with their droves of cattle, and river boatmen crowded the narrow streets. There were opportunities on every hand. It was reconstruction time. The Civil War was over. Louis Bauer arrived in St. Louis.

Trained by German masters, he was a natural choice for the new chair of surgery at the St. Louis College of Physicians and Surgeons, which he had been invited to fill. The town was peopled by strong-willed, dynamic German immigrants who understood Bauer's driving mind. Like Bauer, many of them were dissenters who had been imprisoned for treason in Pomerania in 1848. But let us flip back the calendar!

In Stettin, provincial capital of Pomerania, Louis Bauer was born in July, 1814, of good German burgher parents. Their economic status enabled them to give him an excellent education. In 1833 he became a student of medicine, attending successfully the Universities of Griefswald, Breslau and Berlin and in 1838 passing the state examination as a "physician, surgeon, obstetrician" and in forensic medicine. Almost immediately he became active in politics.

Private practice attracted him to the island of Rügen in the Baltic Sea. Connected by a dam with Stralsund on the mainland, this strategic island of 358 square miles and a goodly population had been fought over for nearly a thousand years. In 1325 it passed from Denmark to Pomerania and in 1815 was conquered by Prussia. Today the Soviet Union has swallowed it whole. Louis Bauer was appointed medical officer of Kreiss County by the Prussian Crown. Turbulent politics engaged his attention as a natural outlet for his vigorous personality. Certain things seem to have occurred so that, as later he was wont to explain, he was transferred at his own request to Stolpe, in Pomerania, where events finally caught up with him.

As a democrat, Bauer became an active party leader in the revolutionary movement and was elected to the lower branch of the Prussian Legislature. He continued to practice, but his opposition to the Crown caused his arrest and imprisonment in 1848 for treason. The revolution persisted, but after 10 weeks Bauer was released; some believe so that the police might shadow him and thus lead them to bigger fish. At any rate, he was about to be rearrested shortly afterward, when word was passed to him of his danger, and he departed by the underground for England in 1849. Soon he established himself, passed the examination of the Royal College of Surgeons and in 1852 became a member of the London Medical Society. Restlessness still dogged him.

Good health and a rousing intellectual curiosity, ingredients of leadership, had survived his brush with the Prussian police. But, somehow, in the transition a frustrating element entered his personality. Perhaps the English were not sufficiently impressed with

* Associate Clinical Professor of Orthopaedics and Lecturer in Anatomy, Yale University, School of Medicine.



Louis Bauer

the young revolutionary's talents. In any event, in 1852 he published a critical report on the technic used by English physicians. This may have raised the ire of his newly found colleagues because the next year, 1853, we find him living in Brooklyn, New York. Friends had preceded him to the United States, and perhaps they had encouraged him to cast his lot with them. Apparently the stiff, unfamiliar atmosphere of English medicine was not to his liking. Brooklyn seemed to be made to order.

Moving from place to place, maturing as he went, always tinged with a sense of superiority, yet dominated on occasion by frustrations, his restless nature finally found peace. His days of medical truancy seemed at an end. To an extraordinary degree not generally appreciated, Bauer proceeded to become one of the great orthopaedic pioneers of the New World. He had had enough of revolutions.

Louis Bauer was one of the founders of Long Island College Hospital, to which he soon was appointed a Professor of Anatomy and Surgery. In 1854, together with Richard Barthelmess, he organized the first private institution devoted to the treatment of orthopaedic disorders in the United States. We know of his penetrating analysis of hereditary diseases and anomalies and of his meticulous interest in their treatment through the *Outline of the Principles and Practice Adopted in the Orthopaedic Institution in Brooklyn*. He and Barthelmess issued this small monograph, dedicating it "To the Medical Profession of the U. S." and calling upon physicians to patronize their new institution.

These intrepid organizers and first-class orthopaedists describe their qualifications and methods, asserting that their "care of the crippled is based on a strictly scientific plan, embracing all those auxiliary means which the advancement of modern surgery affords." They assure their colleagues that "all mystery is repudiated and everyone is invited to call and inspect the institute." Students were solicited on Tuesdays and Fridays from 3 to 6 P.M. for free lectures. Actually, Bauer compliments the English teaching system by imitating it in his own institution.

He ably described many deformities and "traced their causes to material alteration within the great nervous centres, manifesting themselves either as increased or diminished enervation. The former by contractions of vigor, the latter by paralysis." Bauer recommended myotomies or tenotomies under chloroform. As an exponent of rest, he emphasized "proper positioning of limbs at rest so, if anchylosis should ensue, the best functional position would become usable." He advocated incision and drainage of joints, closed reduction of congenitally dislocated hips and special exercises for scoliosis "not to be given by every dancing master or quack" but by one especially skillful in their

ways. His descriptions and basic ideas concerning scoliosis, omitting spinal fusion, are as true today as they were then. For example, he advocated "counter pressure upon the most projecting parts of the spine as a new mode of extension," ridiculing Taylor, his New York contemporary, for his bracing. This is echoed in our turnbuckle and segmental jackets and localizer plasters of modern orthopaedics.

Furthermore, Bauer reports the first statistical study of idiopathic scoliosis, a name he also uses. He found "99 left and 647 right lateral dorsal rotary types." The "young, feeble, anaemic females at the puberty period, whose blood was not in a good state" were more susceptible. The basic disturbance was said to be located within the intervertebral fibrocartilage, and myotomies were fruitless as corrective procedures. Chiefly "rest in the horizontal position for long periods" plus his counterpressure and a spinal support were his approved methods. His institute was approved by the prominent New York physicians Sayre, of Bellevue Hospital, and Parker, of Physicians and Surgeons College.

Not to overreport this early enterprise, but because of its originality one more item seems to be important for our understanding of his environment and times. He is very careful to state that "the institution is located in a fashionable and most healthy part of Brooklyn; as a matter of fact the sexes are separated, young ladies under the care of Mrs. Barthelmess."

In the same year, 1854, he reports on a critical examination of a pathologic specimen of softening of the intervertebral fibrocartilage in a case of Dr. Sayre. He indicates a possible protrusion as a basic disorder. Soon Bauer was made Health Officer of the City of Brooklyn and served 2 years. Never again did he evince an interest in politics; instead, he published many penetrating medical articles, including a textbook of orthopaedic surgery in German, 1870

16 years of practice and teaching in the metropolitan area brought him considerable renown, and he was a frequent guest lecturer. Some of his ideas are incorporated in a monograph of lectures given at McGill University Medical College, Montreal, in 1868. He complains about "those who have stolen *his* ideas and not given any credit to *him*." Such paranoia seems to be part and parcel of many scientific leaders of his day, and even of *our* times.

Ideas are like perennial plants: living for a short while, they seem to die, then to revive. Bauer's concepts of causes and treatments were revolutionary in his day, but now they have been rediscovered and are regarded as "new" and revealing. Actually, only correlations are involved, most of which wax and wane as other scientific weapons are brought to bear on orthopaedic disorders. He knew nothing of the yet unborn science of genetics in relation to hereditary disorders, yet he correlated trauma with osteochondroses of youth and described them as familial in some instances. He reasoned that the ligamentum teres with its vessels might be crushed in the hip joint and a coxal synovitis ensue. For resting the hip joint he designed a shell of leather for the body and the extremities, with feet connected and held in a neutral position.

For all that, his results still are reported by him with a touch of mystery. He indicates a case of paralysis that was treated unfavorably by the great Duchenne, then handled successfully by himself "through the proper use of the electric current," a modality resorted to frequently in his practice.

Bauer's really big moment came when, in 1869, the German burghers of St. Louis called him to their frontier metropolis. He was appointed professor of surgery at the St. Louis College of Physicians and Surgeons, consulting surgeon to the City Hospital, and later became the dean. Finally he came into his own among the people whom he loved and who understood him. Formerly he bore his lot fretfully, as his writings indicate. St. Louis was a haven for

him. His capacities for leadership were given full play while he served the university and the community with honor. There are no more bursts of paranoia; instead, a regular stream of valuable material flowed from his pen, beginning with *Contributions to Medicine and Surgery*, published in 1870. The war was over, for both the people and Bauer the orthopaedist. His real worth was understood by his St. Louisians.

Sir Arthur Keith in his classic *Menders of the Maimed*, published in 1919, quotes no less a critic than the immortal Thomas, of Liverpool, who says of Louis Bauer, "In my early life, I was in my practice a close imitator and an ardent admirer of my friend, Dr. Louis Bauer, the best exponent of American orthopedics." Keith proceeds to give his own appraisal of him, "Everything we can now learn of this American pioneer justifies the acumen of Thomas's earlier judgment." Sir Arthur rated this high opinion on Bauer's concepts of the mechanism of the living, human body. It would seem that Bauer had spent much time studying Marshall Hall, Delpech and Duchenne relative to the action of muscles, nerves and reflex centers. To this Bauer added his own personal observations and applied them with success to the treatment of deformities that are produced by irregular, in-co-ordinated, muscular action. He was particularly critical of most methods of applied traction to the lower extremities. He insisted that traction never was able to separate surfaces of the inflamed joint. However, it must be said that he did overlook, as Keith so fittingly states, "the peculiar virtue of the method of traction applied by Davis in 1860." It would seem that Davis, in New York, and Thomas, in Liverpool, independently had come upon the same idea relative to splinting the knee. This was the forerunner and led eventually to the classic Thomas splint, which immobilized so well, and still does today, the entire lower extremity. Bauer insisted that the splint, which supposedly was immobilizing the extremity, should not be fastened to the foot of the bed. Attaching a movable patient to an immovable bed seemed to him to be a foolish principle.

Bauer also insisted upon rest in the beginning and throughout the entire span of time required in the treatment of tuberculosis of the vertebrae. Fresh air, sunshine and movement were regarded by him with disgust. Any kind of mechanical apparatus, in particular "the spinal assistant" of Charles Fayette Taylor, was regarded by him as "make believe," because it did not relieve the spinal column of its weight-bearing function. He held to the belief that it was impossible to immobilize the spine except when resting flat on the bed.

During his 12 years of occupancy of the chair in surgery, Bauer seems to have been very happy. His published observations and principles were read widely by orthopaedists. He retired with an honorable reputation, working in a senior consulting capacity until 1898. Today a place in our specialty's Hall of Fame is ensured.

REFERENCES

Bauer, Louis: *Contributions to Medicine and Surgery*, St. Louis, Studley, 1870.

———: A critical examination of a pathological specimen of softening of the intervertebral fibro-cartilages, New York J. Med., 1854.

———: Handbuch der orthopädeschen Chirurgie, tr. by B L. Scharlau, Berlin, Hirschwald, 1870.

———: Monograph of Lectures on Causes, Pathology and Treatment of Joint Diseases *delivered to students and others at* McGill University Medical College, Montreal, Canada, 1868.

Bauer, Louis, and Barthelmess, Richard: Outlines of the *Principles and Practice Adopted* in the Orthopaedic Institution of Brooklyn, New York, Schmidt, 1854.

Keith, Sir Arthur: Menders of the Maimed (Limited Ed.), pp. 175-177, *Philadelphia*, Lippincott, 1951.

Medical and Surgical Registry of the United States and Canada, ed. 5, Detroit and Chicago, Polk, 1898.

The National Cyclopaedia of American Biography, Being the History of the United States, New York, White, 5:482, 1894.

2

Re-evaluation of Etiologic Factors in Congenital Anomalies of the Skeleton

THEODORE H. VINKE, M.D.*

In recent years many methods have been described that permit experimental production of congenital skeletal anomalies. Experiments concerned with dietary procedures have been performed for many years and pursued most actively. Prenatal exposure of the mother to large doses of roentgen rays or to toxic dyes, such as trypan blue, also lead to skeletal anomalies. In addition, injection of hormones, such as insulin or cortisone, into the pregnant female can result in certain skeletal anomalies. However, most investigators in this field are of the opinion that human mothers are exposed only rarely during pregnancy to conditions comparable with those of these experiments. It must be kept in mind that genetic factors enter into the formation of congenital defects in man and in animal.

Warkany and Nelson[14] have shown that in rats under strict experimental conditions, diets deficient in riboflavin cause congenital deformities with an established pattern. Under the best conditions, about one third of the young born to deficient mothers are found to be normal. Many of the abnormal young can be recognized as such by external inspection. They show shortening of the mandible and the extremities, protrusion of the tongue and various degrees of syndactylism. Other abnormalities are recognized after the specimens are cleared. There may be shortening of the radius, the ulna, the tibia and the fibula and fused ribs. Inspection of the mouth reveals a posterior cleft palate in about one half of the abnormal young.

In recent studies Warkany and Deuschle[13] analyzed the dentofacial changes induced in these rats and found definite dependence of dental differentiation from the skeletal development. It is interesting to note that some bones are affected more than others. The cranium, the vertebral column, the scapula and the pelvic bones are involved hardly at all. Histologic sections[10] show that an irregular retarded ossification, with malformations, is present in the specimens. In mildly abnormal tibias the metaphyseal lines of ossification take an irregular and somewhat wavy course. Large cartilaginous areas remain enclosed within the diaphysis. Sections of the paws also show irregular and retarded ossifications. Many abnormal bones are noted: some are shortened, and others are lacking. Some bones appear to be fused. Others are wider than normal, and some of the interosseous spaces and articulations are not developed. If the diet is supplemented in rats before the 13th day of gestation, it prevents all skeletal malformations. According to Warkany, these findings suggest that riboflavin is needed for the development of the skeleton before the 13th and the 14th days of embryogenesis. After the 13th and the 14th days of gestation this

*Cincinnati, Ohio.

deficiency does not interfere seriously with the general growth and the increased weight of the rat. Such dietary experiments are difficult, and the yield of abnormal young may be very small, unless special breeding technics are used. Warkany has pointed out that many forms of reproductive failure may be caused by maternal vitamin deficiencies. Sterility, fetal death, resorption or abortion, neonatal weakness or death of the offspring probably is the most frequent result of practical importance in animal, as well as in human, reproduction.

"Malformations" were omitted from the list, since they can occur at any stage of development and play a part in many forms of reproductive failure. Under adverse conditions few deformed fetuses reach the natal period, and, therefore, "congenital" malformations are relatively rare manifestations of maternal dietary deficiency. They occur in exceptional cases as a result of *borderline* deficiencies, which injure the developing embryo but permit continuation of pregnancy. As a rule, mitigation of a borderline deficiency leads to the birth of normal young, but further deprivation leads to embryonic death. Therefore, it is difficult to obtain congenital malformations of dietary origin, even under experimental conditions. This should be kept in mind before hasty conclusions are drawn as to the origin of human malformations. It is interesting that vitamin B deficiencies, other than of riboflavin, have been investigated, and some are found to be capable of producing malformations. Hogan[6] and co-workers found hydrocephalus in rats attributable to *vitamin* B_{12} deficiency. Nelson, Asling and Evans[12] produced a folic acid deficiency in pregnant rats by a combination of a deficient diet and a folic acid antagonist. The malformations produced with these deficiencies of the B group resemble in many respects those produced by riboflavin, which also is a member of the B group of vitamins

It is of interest in this connection that *vitamin A* deficiency of the pregnant rat results in malformations entirely different from those of the B deficiencies.

Warkany[15] has shown that it is possible to produce congenital malformations in rats by maternal *vitamin A* deficiency: the eye is the organ affected most frequently in the syndrome. Malformations of the genital tract follow next in frequency. If depleted mothers receive a single large dose of *vitamin A* on different days, some on the 10th, others on the 11th, the 12th, the 13th, the 14th and the 15th days of gestation, it is found that the percentage of abnormal young becomes smaller the earlier the supplement is given, and that the syndrome of malformation can be modified.

Mellanby[10] concludes that the general function of *vitamin A* aids in controlling the shape of growing bone by influencing the position and the activity of osteoclasts and osteoblasts. He has found that vitamin deficiency causes a general thickening and dysplasia of bone by its effect on osteoclasts and osteoblasts. All the bones of the body are involved to a varying degree. Mellanby also has shown repeatedly that *vitamin A* deficiency in growing animals causes widespread nerve degeneration, both central and peripheral. However, these experiments refer to changes elicited in animals after birth, and not to congenital defects.

It may be said here again that it is unlikely that human congenital malformations *can be attributed to maternal dietary deficiencies*. In populations whose food supply is restricted by war, custom or imprisonment, conditions can arise that are somewhat comparable with these experiments. Thus, endemic cretinism is caused by maternal iodine deficiency, but still it has not been proved whether or not other congenital anomalies are induced in the human being under certain comparable abnormal conditions.

Congenital bowing of the bones of the forearms and the legs is seen in rats when their mothers are fed a diet deficient in *vitamin D* and there is an abnormally high calcium

phosphorus ratio. These deformities can be prevented by the addition of *vitamin D* to the maternal diet.

In addition to dietary deficiencies, positive teratogenic factors have been employed to induce congenital malformations.

Exposure of the mother to large doses of roentgen rays has resulted in congenital malformations. Wilson and Karr[17] demonstrated that rat embryos, which were irradiated, altered greatly the rate of intra-uterine mortality, as well as the incidence of malformations in the brain and the skull. Warkany and Schraffenberger[16] were able to establish very definite patterns of skeletal deformities in newborn rats as being characteristic of irradiation on specific days of gestation. It is of interest that by irradiation of pregnant rats on about the 13th or the 14th day of pregnancy, skeletal malformations similar to those of riboflavin deficiencies are induced. Included in these malformations are shortness of the lower jaw, protrusion of the tongue, cleft palate, shortness of the extremities, clubfeet and syndactylism. However, the roentgen syndrome is more varied than that produced by the riboflavin deficiencies.

Halsey J. Baggs[1] exposed mice to roentgen rays and found that there was a translocation of chromosome particles carrying genes; that an intertranslocated gene, with its neighbors, produced the anomaly of clubfeet, and, by inbreeding, carried this anomaly through hundreds of mice. According to the work of Baggs, the earliest foot defect is associated with the formation of a blisterlike bleb that raises the epithelium of the foot, usually in the localized area. This condition usually is found between the 12th and the 15th days of prenatal life and is followed by the escape of blood into the bleb and the formation of a localized blood clot. These blood clots may persist until birth. Baggs states that the extent of the pathologic processes and the anatomic location determine whether or not the developing foot will show congenital amputations, clubbing, etc.

There appears to be some agreement as to the period of time that may be considered to be critical, in which the prenatal tissue is affected easily. This period is not the same for all agents for all organs. It is also important to note that one cannot ascertain from a malformation the time at which the injurious agent acts. It is possible to state that a malformation may have occurred before a certain time, but how much before this critical period the injurious agent acted it is difficult to establish.

Chemicals, such as alcohol, benzine, nicotine, quinine, lead, mercury and iodine have been suspected to be injurious agents, but experimental studies have been rather contradictory.

Hormonal disturbances of the mother must be considered a potential danger to the fetus. Fraser and co-workers[4] showed that injection of cortisone into pregnant mice of a certain strain induced cleft palate in the young.

There also is an increase in congenital anomalies in infants born of diabetic mothers. The inadequately treated mothers gave birth to children with a high mortality rate.

Much work has been done on the development of the chick under insulin treatment. Landauer[9] has analyzed in many careful studies the effect of this hormone on the chemical processes of prenatal development. Duraiswani's[3] experiments probably are better known to orthopaedists. In his experiments some of the "chicks" present roentgenologic appearances resembling osteogenesis imperfecta, achondroplasia and osteochondrodystrophy. Other abnormalities found are spina bifida, partial or complete suppression of development of one or more vertebrae, scoliosis, clubfeet, arthrogryposis, bowing of the tibia and dislocation of hips and knees. A variety of abnormalities has been produced by using Synthalin, thalium salts, sulfonamides and cortisone. Insulin-produced deformities can be prevented with nicotinamide and riboflavin.

Deformities and embryonic malformations

have been produced by submitting pregnant rats to anoxia inside a bell jar and also by injecting eggs with tetanus toxin. Of course, we recognize that the experiments with chicken eggs may not be entirely comparable with those in mammals, but certainly they are significant.

These experiments show clearly that environmental factors, acting during pregnancy, can induce congenital malformations. There is no evidence that any of these malformations are hereditary. They must be distinguished clearly from malformations that are determined genetically and hereditary, according to mendelian transmission. Such hereditary anomalies originate by spontaneous mutation or they can be induced by roentgen-ray effects on the germ cells.

Differentiation must be made as to those deformities of genetic origin and those produced experimentally as nonhereditary. For instance, cleft palate and syndactylism may be of genetic origin. Similar deformities can be produced experimentally by certain vitamin-deficient diets. Thus, genetic and environmental factors may lead to the same kind of congenital malformations. Identical malformations may be hereditary in one case and nonhereditary in another. In man, certain malformations are inherited as a dominant trait, such as complete absence of all extremities or absence of one extremity. Absence of the humerus and the ulna is rare. Absence of the radius occurs much more often and is associated with club hand. Absence of the fibula is encountered more frequently than that of the femur or the tibia. Of course, clubfoot is one of the most frequent deformities of the foot. Supernumerary fingers or toes may be found in a single member of a sibship, and there are pedigrees in which polydactylia is inherited as a dominant trait and often is associated with other malformations. Cleidocranial dysostosis is a congenital syndrome; it is characterized by the absence of the clavicle and a delayed ossification of the skull, and is transmitted as a dominant character. A congenital webbed neck also is combined with edema of hands and feet. Chondrodystrophy is caused by disorder of the cartilage, which begins in prenatal life. Advanced maternal age has been considered as a possible etiologic factor in this malformation, in which males and females are affected equally. The basic pathologic process is disturbance of the enchondral ossification, caused by inability of the epiphyseal plate to produce a sufficient number of columnar cartilage cells. Multiple exostoses are hereditary in the majority of instances. Fibrodysplasia of the bone may affect one or more bones of the skeleton.

In arthrogryposis there is a contracture of joints in flexion. The term has been used in a functional sense of a carpopedal spasm and also for congenital abnormalities of the constituents of the joint. In arthrogryposis, dislocations of the hip and other joints occur frequently. This syndrome originates early in embryonic life, before intra-uterine pressure becomes an important factor.

Murk Jansen[7] thought that the problems of malformation were explained on a mechanical basis. He looked for mechanical malformations due to pressures that acted on the fetus, such as various sizes of the amnion and differences in hydrostatic pressure in the uterus.

Children may be deformed by a prenatal infectious disease of the mother. In congenital defects following maternal rubella, Gregg[5] showed that many cases of congenital cataracts, associated with cardiac or other lesions, occurred in those giving a history of maternal rubella during pregnancy. Congenital deformities associated with these conditions were cataracts, cardiac deformities, deafness, mental retardation, clubfeet and dental problems. Although it was thought at first that 100 per cent of the mothers who contracted rubella during the first trimester of pregnancy gave birth to infants with congenital anomalies, it now is thought that this incidence may be much smaller.

According to an article by Kay, Rossner

and Stein,[8] a series of 154 cases of virus disease in early pregnancy other than rubella show that 21 fetal anomalies occurred. These findings raise the question as to whether or not such malformations may be preventable. However, as yet there is not sufficient information available on which to base a program designed to prevent all congenital anomalies in man. Further studies of causes of immaturity, stillbirth and congenital anomalies of various types are required. As in the case in other diseases, prenatal conditions may be observed through case study and experimentation. However, human beings cannot be subjected to the same conditions as are experimental animals. Consequently, broad conclusions cannot be drawn from experiments.

In experimental animals we see certain patterns of anomalies following certain treatments. Therefore, we must look for such patterns in congenital anomalies in man. Perhaps we shall be able to recognize some syndromes by associated minor abnormalities. Supernumerary teeth in cases of cleft palate is one such condition. In orthopaedic surgery we find frequent mention of "café au lait" markings and other evidences of neurofibromatosis associated with other congenital lesions. Beveridge Moore[11] reported that in 95 per cent of the cases of congenital anomalies, associated pathologic changes in peripheral nerves were present It is known that congenital lesions of minimal degree are not recognized, but by careful examination they will be found with associated patterns of maldevelopment. Bechtol and Mossman[2] have suggested that abnormal development of the skeletal muscle is associated with clubfoot.

These examples indicate that it is possible for the clinician to learn a great deal from animal experiments However, he must not assume hastily that the difficult and complex experiments that result in malformations in animals imitate prenatal human conditions The causes of malformations of children must be investigated also by clinical observations of the patient, his environment and his pedigree.

REFERENCES

1. Bagg, Halsey J.: Hereditary abnormalities of limbs, their origin and transmission with reference to descendants of x-rayed mice, Am. J. Anat. 43:167, 1929.
2. Bechtol, C. O., and Mossman, H. W.: Club foot; an embryological study of associated muscle anomalies, J. Bone & Joint Surg. 32-A:827-836, 1950.
3. Duraiswani, P. K.: Experimental causation of congenital skeletal defects and its significance in orthopaedic surgery, J. Bone & Joint Surg. 34-B:646-698, 1952.
4. Fraser, F. C., and Fainstat, T. D.: Production of congenital defects in the offspring of pregnant mice treated with cortisone, Pediatrics 8:527-533, 1951.
5. Gregg, N. M.: Rubella during pregnancy of the mother, with its sequelae of congenital defects in the child, M. J. Australia 1:313, 1945.
6. Hogan *et al:* Vitamin B_{12} a factor in prevention of hydrocephalus in infant rats, Proc. Soc. Exper. Biol. & Med. 76:349-353, 1951.
7. Jansen, Murk: Achondroplasia, Its Nature and Its Cause, London, Baillière, Tindall & Cox, 1912.
8. Kay, M. K., Rossner, D. C., and Stein, I. F.: Viral diseases in pregnancy and their effect upon the embryo and fetus, Am. J. Obst. & Gynec. 65:109-119, 1953.
9. Landauer, W., and Rhodes, M. B.: Further observations on the teratogenic nature of insulin and its modification by supplementary treatment, J. Exper. Zool. 119: 221-262, 1952.
10. Mellanby, E.: Vitamin A and bone growth: the reversibility of vitamin A deficiency changes, J. Physiol. 105:382-397, 1947.
11. Moore, Beveridge H.: Some orthopaedic relationships of neurofibromatosis, J. Bone & Joint Surg. 23:109-140, 1941.
12. Nelson, M. M., Asling, C. W., and Evans, H. M.: Congenital abnormalities in fetal rats resulting from pteroylglutamic "folic" acid deficiency during gestation, Anat. Rec. 106:309, 1950.
13. Warkany, J , and Deuschle, F. M.: Congenital malformations induced in rats by maternal riboflavin deficiency: dentofacial changes, J. A. D. A. 51:138-154, 1955.

14. Warkany, J., and Nelson, R. C.: Skeletal abnormalities in the offspring of rats reared on deficient diets, Anat. Rec. 79: 83-100, 1941; Skeletal abnormalities induced in rats by maternal nutritional deficiency, Arch. Path. 34:375-384, 1942.
15. Warkany, J., and Schraffenberger, E.: Congenital malformations induced in rats by maternal nutritional deficiency: VI. The preventive factor, J. Nutrition 27: 477-484, 1944.
16. ———: Congenital malformations induced in rats by roentgen rays, Am. J. Roentgenol. 57:455-463, 1947.
17. Wilson, James G., and Karr, John W.: Effects of irradiation on embryonic development, Am. J. Anat. 88:1-33, 1951.

Re-evaluation de Factores Etiologic in Congenite Anomalias del Skeleto

Summario in Interlingua

Es discutite le production experimental de congenite anomalias per medio de varie technicas, i.a. le uso de colorantes toxic, de hormones, e de dietas a deficientias vitaminic. Attention special es prestate al labores de Warkany con dietas a deficientia del materne vitamina A e de riboflavina. Es presentate le methodo de exponer animales pregnante a radios X e etiam le labores de Duraiswani in le disveloppamento de gallinettas sub tractamento a insulina. Es formulate le conclusion que factores genetic e ambiental pote effectuar le mesme typo de deformitates congenite. Un mesme malformation pote esser hereditari in un caso e non-hereditari in un altere. In ultra de deficientias dietari del matre, etiam infectiones prenatal pote causar deformitates Un exemplo es rubella materne que causa cataractas, deformitates cardiac, retardation mental, etc. In animales experimental nos observa certe configurationes de anomalias occurrente post certe typos de tractamento. Per consequente nos pote expectar que le anomalias congenite in humanos se presenta in simile configurationes. Forsan nos pote recognoscer certe syndromes super le base de minor anormalitates associate. In iste campo le clinico pote apprender multo ab experimentos animal. Nonobstante, nos non debe concluder precipitatemente que le complexe e difficile experimentos que resulta in malformationes in animales imita le conditiones prenatal in humanos. Le causa de malformation in infantes human debe esser investigate per le observation clinic del patiente e per le studio de su milieu e su arbore genealogic.

3

Environmental Causes of Abnormal Embryonic Development

PETER GRUENWALD, M.D.*

The treatment of congenital malformations is an unsatisfactory undertaking and seldom results in restoring normal function and appearance. Therefore, it is particularly desirable to prevent the development of malformations. In order to do this, we must know the causes of abnormal development, and particularly those originating in the environment, which may be affected by preventive measures. These environmental causes (as opposed to genetic ones) include mechanical force, chemical alterations (excess or deficiency of substances), radiation, abnormal temperature, infection and, in man and mammals, influences of an altered maternal environment. The effect of each of these factors has been demonstrated excellently in experiments. Some environmental agents affect more easily or exclusively embryos of lower animals developing outside the maternal organism; these are only of basic interest in connection with human malformations.

MECHANICAL AGENTS

Mechanical agents affect embryos unprotected by the maternal organism. It is known that the shaking or the jarring of eggs produces malformations in the trout and in the chick; this hazard is taken into account in commercial hatching procedures. In mammals, surgical procedures have been performed on embryos *in utero*. The greatest difficulty in such experiments lies in the tendency toward abortion whenever uterus and embryos have been tampered with. This same factor accounts for the extreme rarity of well-authenticated human cases in which mechanical injury (such as an accident or an attempted abortion) has been followed by further development and malformations.

Some writers have considered the so-called intra-uterine amputation of portions of limbs and similar defects to be due to mechanical injury by amniotic bands. However, it is likely that there is an underlying disease condition of the skin and even of deep tissues.[28]

CHEMICAL ALTERATIONS

Chemical effects on development may be due to the introduction of substances foreign to the organism (poisons) or to abnormal concentration, high or low, of normal constituents (e.g., hormones, vitamins). It is easy, particularly in lower animals, to produce a great variety of malformations by introducing chemicals into the medium in which the embryos develop.[2] Of some interest are the effects of several substances on the skeleton and the limbs of chick embryos. Achondroplasia, of which 6 hereditary forms are known in the chick, also can be produced by deficiency of manganese, riboflavin, lactic or nicotinic acid. Less characteristic forms of micromelia result from

*Director of Laboratories, Margaret Hague Maternity Hospital, Jersey City, New Jersey.

selenium poisoning or localized treatment with colchicine. Defects of the distal portions of the vertebral column, which occur in chicks sporadically as well as by the action of several hereditary traits, may be produced by injection of insulin into the egg. For a review of this subject the reader is referred to Landauer.[18]

POISONS

Very little successful work on the effect of poisons on mammalian embryos is on record. Apparently, here, as in mechanical injuries, it is very difficult to find a level of toxicity that will affect the developing embryos and not interfere with continuation of pregnancy. Yet, it is possible that such conditions exist. We have learned from the study of the effects of virus diseases on human embryos (see below) that there is no parallelism in the severity of the effects on mother and embryo. One example from the field of experimentation is the production of various malformations by injection of trypan blue into rats before and during pregnancy.[8] Lead poisoning, asymptomatic in the mother, has been suspected of producing malformations in man,[13] but no conclusive information yet has been obtained. Iodine deficiency may lead to cretinism with its well-known disturbances of mental and physical development.

VITAMINS

Abnormal levels, mostly deficiencies, of vitamins have been studied experimentally as to their effects on the embryo. Antimetabolites have been used with excellent success in some of the more recent work in this field. Numerous investigators have reported the changes induced in laboratory animals by deficiencies of vitamin A,[33] riboflavin,[32] folic acid,[10] pantothenic acid,[9] linoleic acid[21] and vitamin D.[31] Again, there is a narrow range in which deficiencies produce developmental changes without interfering with continued pregnancy. For this reason, only very few clear-cut cases of intra-uterine changes due to vitamin deficiencies have been reported in man. Low grade and uncharacteristic deficiencies of the fetus probably are more common, but these cannot be apprehended. Estimates of the effects of poor nutrition range up to the statement of Burke and coworkers,[5] to the effect that nearly all perinatal deaths, the majority of malformations and all instances of prematurity occurred in their series in mothers with inadequate diet. This is to be viewed with suspicion, in view of so many other known causes of abnormalities. In many vitamin deficiencies of experimental animals, skeletal changes, not necessarily similar to those in children or adults, are prominent.

HORMONAL IMBALANCES

Hormonal imbalances have been produced experimentally by the administration of hormones to pregnant animals and, in rare instances, by the surgical removal of endocrine organs from embryos. A large volume of literature has accumulated on the effect of these procedures upon the genital organs Of interest are the effects that Jost attributed to the administration of ACTH to rat embryos and later recognized to be due to pressor substances.[16] These produced necrosis and loss of the distal portions of the extremities and the tail of the embryos, resembling in the final outcome some of the so-called amniotic amputations. Cortisone administered to pregnant rats produces cleft palate, varying in incidence in different strains of animals.[7] So far, experiences with women receiving cortisone treatment during pregnancy have not indicated untoward effects on the offspring.[7] According to Langman and van Faassen,[20] thyroid deficiency of pregnant rats produces malformations of the eyes, cleft palate and abnormalities of the skeleton and the extremities

OXYGEN PRESSURE

Variations in oxygen pressure have significant effects on development. Examples of numerous investigations are those of Nau-

joks[22] on chick embryos, describing malformations of the eyes, central nervous system and extremities, and those of Ingalls and coworkers[14] on pregnant mice, in which the embryos showed defects of the eyes, the central nervous system, the palate and other parts, in addition to resorption of embryos. It is not known to what extent anoxia is teratogenic in human beings. Difficulties of human propagation at high altitudes are on record.[15]

RADIATION

Interest in the effects of radiation on embryonic development has increased in recent years, along with other fields concerned with ionizing radiations. The proceedings of a symposium on this subject[23] attest to this and should be consulted for discussion of various basic aspects of the field. Most of the work has been done with roentgen rays, which are relatively easy to handle and control. A number of workers have used closely timed exposure of rat or mouse embryos to determine patterns of abnormal development and their changes as development proceeds. A large variety of skeletal changes feature prominently in the results of this work. The original reports must be consulted for details.

It is essential to distinguish the effects of damage produced by ionizing radiations in the tissues of a developing embryo from a possible influence on the genetic constitution The former results in abnormalities of the individual subjected to radiation; its offspring will not be affected. The latter does not alter the individual in a manner discernible by any means of investigation; however, its offspring may be abnormal This latter abnormality may appear several generations later, and not in a characteristic form, since radiations probably increase the mutation *rate* without producing any mutations that would differ qualitatively from spontaneous ones Therefore, from the experience gathered during one observer's lifetime, it is a gross error to state that irradiation of the gonads does not produce genetic alterations.

TEMPERATURE

So far as we know, direct effects of temperature changes on the embryo are irrelevant in man. Stress of the mother produced by extreme temperature changes might conceivably affect the embryo by the endocrine consequences in the mother. However, this has not been demonstrated so far in human cases. In chick embryos, reduced temperature during certain phases of development has been shown to interfere with normal development[19] or to modify the expression of hereditary malformations.[20]

INFECTION

To what extent infection causes abnormal development is not known. Even the incidence of malformations following rubella during the first trimester of pregnancy is under discussion. More and more investigators suspect that the figures should not be set as high as was believed immediately following the discovery of this relationship. Other viral infections may be involved less frequently. It is not certain that immunization of women will prevent subsequent effects of viral infection on the embryo, since prevention of disease in the mother does not necessarily mean prevention of viremia and infection of the embryo.[21]

Bacterial and protozoan infections may produce severe disease in the fetus (e.g., syphilis, toxoplasmosis), but developmental processes are not involved predominantly.

Several points stand out in the story of virus-induced human malformations as we know it today. One is that here, as in other instances mentioned above, mild and sometimes subclinical diseases of the mother have more profound effects on the embryo than has severe illness. Another point of interest is the question of the nature of the agent versus the time of action. Some observers have stated that rubella during the second month produces changes different from

those arising during the third month, etc, and it was concluded that the action of rubella was a nonspecific one, and that the type of abnormality was determined by the time of action. Against this we must hold the characteristic and limited range of effects of rubella on the embryo. This can be accounted for by a *specific* action of the disease, modified only to a slight extent (within certain limits) by the state of the embryo, and makes it appear likely that the embryo is affected directly and specifically by the virus, rather than in an indirect and nonspecific manner by the maternal disease.

MATERNAL AND UTERINE ENVIRONMENT

The maternal, and more particularly the uterine, environment of the mammalian embryo may have adverse effects on development. This has been demonstrated in purebred strains of mice.[6] Recent studies of pregnancy wastage in women with repeated unsuccessful pregnancies have suggested a significant role of gross uterine deformities or endometrial abnormalities.[1] In mongolism, the high incidence in infants of relatively old women is well known; what kind of maternal factor, uterine or otherwise, is involved has not been determined.

GENETIC AND ENVIRONMENTAL FACTORS

The relationship of genetic and environmental factors is a fundamental problem in both normal and abnormal development. Everyone accepts the fact that normal development is determined or influenced by genetic factors and also by effects of the environment. Yet, it has been difficult to gain acceptance of the same principle in abnormal development In recent years direct evidence has been obtained both for modification of hereditary traits by the environment and for genetic variations in the effect of teratogenic agents The latter has been investigated by Fraser,[7] using the production of cleft palate by cortisone in several purebred strains of mice, and by Landauer,[17] investigating the effects of boric acid on embryos of various strains of chicks.

The morphologically discernible effects of genetic and environmental factors may be quite similar. It has been possible to produce fairly close replicas ("phenocopies") of hereditary abnormalities by altering the environment.[18] In some instances it is known that the basic mechanism in the two similar malformations is different; in other cases one might suspect that essential steps in the basic mechanisms were common to the hereditary trait and its phenocopy. For instance, it is conceivable that an experimental vitamin deficiency and a hereditary metabolic defect influence the same chain of chemical reactions and, therefore, produce similar visible effects. As yet, conclusive proof has not been brought forward.

From the point of view of medical practice, two closely related questions stand out: how can one, faced with a malformation in a patient's offspring, determine the cause of the defect?; and what advice can one give to prevent the occurrence of the same or another abnormality in the future? It must be remembered that reliable methods of genetics and of developmental pathology are based on the planned production of large numbers of offspring under controlled conditions. There are very few hereditary abnormalities in man that are so conspicuous and are transmitted with so little modification in their expression that their genetics can be established even under the adverse conditions of human propagation (e.g., hemophilia). Even smaller is the number of those malformations that can be recognized as caused by a definite environmental agent (e g, rubella, roentgen rays). A number of malformations "run in families," and these are probably, *but not definitely, hereditary.* That leaves the bulk of developmental abnormalities *in the so-called sporadic group;* their cause and genesis are unknown, and speculative explanations have beclouded rather than elucidated the field Even in some of the few instances in which the cause

of a malformation is known, the probability of its occurrence is controversial (e.g., rubella). Some malformations recur repeatedly in some families and are sporadic in the rest of the population. It is extremely difficult to determine whether they are genetically caused in the former event or in all instances, or not at all. The difficulties of the genetic evaluation of human abnormalities have been presented concisely by Snyder.[27] There probably is no other field in pathology in which the gap between well-founded basic knowledge and the lack of its applicability to a given human case is as great as in teratology.

PATHOGENESIS

The pathogenesis of many malformations of animals is well known. Closely spaced embryonic stages have been followed, as in investigations of normal development, and in many instances the abnormal embryos develop their special traits with nearly the same regularity that characterizes normal development. Processes that previously had been postulated have been demonstrated to occur, such as the splitting or the fusion of primordia, or the excess or the retardation of normal development, progressive as well as regressive. Developmental interrelations may add extensive secondary anomalies to a limited primary defect. However, in addition, a type of abnormality that never was envisaged by the fathers of teratology has been shown to be one of the most important ones, i.e., degeneration of previously well-formed parts. It follows hereditary, as well as environmental, causes of maldevelopment. Well-documented hereditary examples reproducing conditions of interest to the orthopaedic surgeon include the nervous system (hereditary ataxia of lambs[3]), muscles (crooked-neck fowl[24]) and extremities (brachydactylia of rabbits[11]) For example, influences of the environment include the effects of selenium compounds on the central nervous system and the extremities of chick embryos.[12] There are indications, both from observation of suitable cases and from comparison with animal material, that the so-called amniotic amputations in man are due similarly to tissue degeneration rather than accidental strangulation by amniotic bands.

In the light of these facts, several teratologic concepts must be reviewed critically. One of these is the *teratogenetische Terminationspunkt* of Schwalbe,[20] which means the latest stage of embryonic development at which a given malformation can develop; another one is the status Bonnevie-Ullrich. The first-mentioned concept stems from the days when teratology developed into a science with a rational basis, largely under the influence of experimental embryology. Developmental processes had been modified in a regular and predictable manner by experimental procedures, and malformations resulted. It was only reasonable to assume that, given adequate knowledge of normal development, one might retrace in one's mind the steps of morphogenesis and thus figure out the point at which development must have deviated to yield a given malformation. This kind of reasoning is based on the occurrence of processes such as retardation, deviation or excess of growth, fusion or splitting of primordia, etc. It is pertinent in certain instances, e.g., transposition of the great vessels, in which it is inconceivable that normal relations of the arteries to the ventricles of the heart could have developed fully and then each artery switch from one ventricle to the other. In another example—tracheo-esophageal fistula combined with atresia of the esophagus—it is equally inconceivable that the normal separation of the trachea from the esophagus could take place and the 2 primordia then reunite by a fistula and one of them divide into 2 portions in a manner as regular as one finds the features of the malformation in question. In these instances it is reasonable and permissible to figure out when the crucial steps in development occur that are modified in a given malformation. The abnormality must have been determined at or before that point. On the other hand,

there are innumerable malformations of the nervous system, the sense organs, the musculoskeletal system or the entire extremities, in which it is likely, and in some instances actually proved, that parts which are defective at birth were laid out and formed normally and then destroyed or deformed by degeneration. In these instances it is impossible to figure out time and mechanism of the abnormal developmental process by examining the individual at birth. Thus it appears that Schwalbe's old concept applies to a limited number of malformations, whereas in others its use is unjustified and misleading.

One of the instances in which entirely unwarranted conclusions have been drawn from the appearances of malformed individuals after birth is the status Bonnevie-Ullrich. Bonnevie[4] described how blebs of cerebrospinal fluid, escaping from the brain and traveling in the subcutaneous tissue, cause disturbances in embryonic tissues in a strain of mice with malformations of the eyes, the extremities and other parts. Ullrich[30] adopted this explanation advanced by Bonnevie for one strain of mice to account for a wide variety of unrelated human malformations. This is unreasonable, not only because, as was pointed out above, the final form of a malformation usually does not reveal the mode of pathogenesis, but also because in large numbers of well-authenticated cases in animals with and without bleb formation the cerebrospinal fluid has not been found to be involved. Actually it is doubtful that Bonnevie's explanation is true, even for that particular strain of mice, as has been pointed out by Jost[16] Moreover, the abnormalities included in the status Bonnevie-Ullrich by various writers are very heterogeneous and should not be considered as one entity. The large number of papers dealing with this alleged status attest to the poor background and judgment of many writers concerning themselves with teratology who pretend to know pathogenetic mechanisms and figure out the time of origin of all malformations, or those who have us believe that many kinds of abnormalities are caused by rampaging cerebrospinal fluid. This retards rather than advances the knowledge of abnormal development.

REFERENCES

1. Aaron, J. B., Levine, W., and Gitman, L.: Obst. & Gynec. 2:461, 1953.
2. Ancel, P.: La Chimiotératogenèse chez les vertébrés, Paris, Doin, 1950.
3. Bogaert, van, L., and Innes, J. R. M.: Arch. Path. 50:36, 1950.
4. Bonnevie, K.: J. Exper. Zool. 67:443, 1934.
5. Burke, B. S., Beal, V. A., Kirkwood, S. B., and Stuart, H. C.: Am. J. Obst. & Gynec. 46:38, 1943.
6. Fekete, E.: Anat. Rec. 98:409, 1947.
7. Fraser, F. C., Kalter, H., Walker, B. E., and Fainstat, T. D.: J. Cell & Comp. Physiol. 43:237, 1954.
8. Gillman, J., Gilbert, C., Gillman, T., and Spence, I.: South African J. M. Sc. 13: 47, 1948.
9. Giroud, A.: Études néonat. 1:5, 1952.
10. Giroud, A., Lefebvres, J., and Dupuis, R.: Rev. internat. vitaminol. 24:420, 1952; Asling, C. W., Nelson, M. M., Wright, H. V., and Evans, H. M.: Anat. Rec. 121:775, 1955; Nelson, M. M., Wright, H. V., Asling, C. W., and Evans, H. M.. J. Nutrition 56:349, 1955.
11. Greene, H. S. N., and Saxton, J. A., Jr.: J. Exper. Med. 69:301, 1939; Inman, O. R.: Anat. Rec. 79:483, 1941.
12. Gruenwald, P.: To be published.
13. Hansmann, G. H., and Perry, M C.: Arch. Path. 30:226, 1940.
14. Ingalls, T. H., Curley, F. J., and Prindle, R. A.: Am. J. Dis. Child. 80:34, 1950.
15. Ingalls, T. H., Prindle, R. A., and Curley, F. J.: New England J. M. 247:758, 1952.
16. Jost, A : Arch. françaises pédiat. 10:865, 1953.
17. Landauer, W.: Genetics 38:216, 1953.
18. ———: Growth Sympos. 12:171, 1948.
19. ———: The hatchability of chicken eggs as influenced by environment and heredity, Storrs Agric. Exper. Stat. Bull. 262, 1951.
20. Langman, J., and Faassen, van, F.: Am. J Ophth 40:65, 1955.
21. Martinet, M.: Ann. méd. 53:286, 1952.

22. Naujoks, H.: Beitr. path. Anat. 113:221, 1953.
23. Oak Ridge National Laboratory, Biology Division: Symposium on effects of radiation and other deleterious agents on embryonic development, J. Cell. & Comp. Physiol., Supp. I, 1954.
24. Rosenberg, L. E.: Anat. Rec. 97:277, 1947.
25. Schick, B.: Acta paediat. 38:563, 1949.
26. Schwalbe, E.: Allgemeine Missbildungslehre (Teratologie) *in* Die Morphologie der Missbildungen des Menschen und der Tiere, Part 1, Jena, Fischer, 1906.
27. Snyder, L. H.: Am. Naturalist 76:129, 1942.
28. Streeter, G. L.: Contrib. Embryol. 22:1, 1930.
29. Sturkie, P. D.: J. Exper. Zool. 93:325, 1943; Landauer, W.: Science 100:553, 1944.
30. Ullrich, O.: Ergebn. inn. Med. 2:412, 1951.
31. Warkany, J.: Am. J. Dis. Child. 66:511, 1943.
32. Warkany, J., and Nelson, R. C.: Anat. Rec. 79:83, 1941; Giroud, A., Lefebvres, J., and Prost, H.: Arch. anat. micr. 42: 41, 1953.
33. Warkany, J., and Schraffenberger, E.: Proc. Soc. Exper. Biol. & Med. 57:49, 1944.

Causas Ambiental de Anormal Disveloppamento Embryonal

Summario in Interlingua

Es passate in revista varie causas ambiental de malformation. Illos include agentes mechanic, effectos de alterationes chimic, radiation, alteration de temperatura, infection, e influentia del organismo materne.

Es discutite le relation inter factores ambiental e genetic. Es notate le similitude de certes de lor effectos e mechanismos. Difficultates incontrate in le interpretation de causa e pathogenese de malformationes human es mentionate, e le fallacia del si-appellate stato Bonnevie-Ullrich es signalate. Le limitationes del deduction de causa e pathogenese de un malformation super le base de su apparition final al nascentia es obvie.

4

The Genetics of Joint Diseases*

ROBERT M. STECHER, M.D.†

Genetics as a possible etiologic factor in various types of joint disease is commanding more and more attention. This is due in part to advances in technical knowledge of the science of genetics and in part to uncertainty concerning the etiology of these diseases. It is the purpose of this chapter to review the evidence and pursue the argument that supports the hereditary basis of joint diseases and to weigh the importance of heredity against other known causes.

In the study of genetics the process is always the same. First, disease or a syndrome is recognized, described and defined. Finally, specific diagnostic features are established, and case reports appear. The familial nature of the disease is confirmed when cases are described in parent and children, in several members of the same sibship and in twins. When these appear in sufficient numbers they are assembled and analyzed *in toto.* Then correction can be made for families of small size; decision can be made as to whether inheritance is as a dominant or a recessive; and computation can be made as to penetrance and whether or not the character is sex linked or sex influenced. If data are available as to incidence in the general population, an estimate can be made as to gene frequency. Regarding arthritis, all these steps have been followed satisfactorily concerning Heberden's nodes, they have been followed partially in rheumatoid arthritis, spondylitis and gout, and heredity is barely suspected in osteoarthritis of the hip.

In hereditary studies it is important that the diagnosis be made accurately and data restricted to the same disease. Former studies have grouped different diseases together as the rheumatic syndrome linking in the same pedigree rheumatic fever, rheumatoid arthritis, spondylitis and osteoarthritis, which has led to inaccurate and confusing conclusions. Even in the same disease, as osteoarthritis, affecting different joints, differentiation must be made. It will be shown that the heredity of osteoarthritis of the fingers, the hips and the spine vary widely from one another.

It is not necessary to have a high family incidence of disease, direct transmission from parent to offspring, or to attain mendelian ratios of involvement to prove that heredity plays a part in the causation of disease. In diseases of the joints inheritance often is irregular, penetrance is incomplete, and involvement is modified by sex, age and the menopause. The late age of onset also adds to the difficulty of analysis, because many susceptible people die before the disease develops. The older the age of onset the more important environmental factors are, and the likelihood of avoiding the disease increases. In other words, joint disease often is multifactorial, and other conditions besides hereditary susceptibility must be fulfilled for the disease to develop. If there is no hereditary susceptibility, the disease does not occur.

*From the Department of Medicine, Cleveland City Hospital, Western Reserve University Medical School

†Associate Clinical Professor of Medicine, Western Reserve University Medical School, Cleveland, Ohio

Among the joint diseases in which heredity seems to play a part, and which will be discussed here, are Heberden's nodes, rheumatic fever, rheumatoid arthritis, ankylosing spondylitis, gout and osteoarthritis of the hip. The joint disease of rheumatic fever is temporary and never permanently deforming. However, in this disease 2 factors, a hereditary susceptibility and an environmental factor or a streptococcal infection, have been identified. Gout is a metabolic disease, but joints are involved and at times damaged seriously. Osteoarthritis of the hips is a complicated disease or a collection of different diseases, and much of the discussion here is probably more philosophical than scientific. However, these are specific diseases, each associated with pathologic lesions that are recognized readily. Nothing will be said about such functional diseases as fibrositis, menopausal arthritis or psychogenic rheumatism.

HEBERDEN'S NODES

Heberden's nodes or osteoarthritis of the finger joints arise in 2 different ways.[18] In men particularly, and after injury, enlargement of the terminal joints appears promptly, develops in a short time to a final stage and remains unchanged throughout life. Only injured joints are affected. This is a true traumatic arthritis and is mentioned only for completeness. Traumatic Heberden's nodes will not be mentioned again.

Our main interest is centered in idiopathic Heberden's nodes, those that arise spontaneously without recognizable cause, usually in women and most often at or about the time of the menopause. This disease starts in 1 finger and spreads in months or years to others. Having seen a family of 1 man and 4 sisters with enlarged fingers, a survey of the population at large with observation of nearly 8,000 individuals gave information about the general clinical appearance of the disease, its distribution among the different fingers, the sex ratios of involvement and the incidence of the disease in each decade of life. With this information available, it was possible to show that Heberden's nodes ran in families, that mothers were affected twice and sisters 3 times as commonly as the general population, and that certain combinations of family involvement were not expected to occur by chance alone more often than once in 10 million and once in 5 million families.[19] Obviously, penetrance in women depended upon age. Gene frequency analysis[20] showed that this disease was sex influenced, being dominant in women and recessive in men. After the 7th decade, when penetrance is complete, the incidence of Heberden's nodes in women is 30 per cent and in men 3 per cent. Being dominant in women, they are affected when they are heterozygous. Men are affected only if they are homozygous. However, the heterozygous men transmit the gene to one half of their progeny. These women develop Heberden's nodes, their brothers become carriers. Men become affected only if they are homozygous. It will be expected that one fourth of these brothers and three fourths of these sisters will be affected. It is expected that the trait will be inherited by all their daughters.

When pedigrees were divided into 2 groups, 1 in which 1 parent was affected and 1 in which no parent was affected, it was found that the 1st group had an appreciably higher proportion of affected children. In the families without affected parents it was assumed that the trait was inherited from the phenotypically normal father who was genotypically heterozygous for Heberden's nodes. In the group of families with an affected mother it is expected, with random mating, that 30 per cent of the fathers will carry the gene and account for the higher incidence of the trait. In families with known double inheritance, as identified by the presence of affected sons, the daughters are expected to be affected in the ratio of 3 to 1. In 4 such families, 8 of 11 daughters were found to be affected, which came very close to the expected ratio.

Certain conditions interfere with the realization of these results. The age of onset is delayed; therefore, it is inevitable that some women will die before the trait develops or is recognized. The development of Heberden's nodes is related in some way to the menopause, because one half of our patients noted Heberden's nodes within 3 years of the last menstrual period.[19] When the menopause preceded the onset by many years, it was found that a hysterectomy had been performed. When the onset preceded the menopause, no ready explanation was found.

In the investigation of a large pedigree involving 30 individuals in 3 generations, several women who theoretically should have had Heberden's nodes did not show them.[21] Lateral roentgenograms of the fingers revealed small nests of calcium deposit in the extensor tendons near their attachment to the distant phalanx. These have been seen in the early stages of the disease, and it was decided that they were manifestations of poor expressivity of the disease.

In the author's experience, Heberden's nodes are a specific type of joint involvement confined to the finger joints, most commonly affecting the terminal joints but also involving proximal joints. They are not part of a generalized osteoarthritic syndrome.[17] None of these patients had severe, generalized or crippling joint disease. It is true that patients with Heberden's nodes complained of creaking of the knees and took aspirin more often than did the control series. This was because, in the course of the studies, patients with Heberden's nodes were seen over and over again, often during a period of 10 years, and they had ample opportunity to complain and to get prescriptions. The control subjects were seen only once; they were surprised and puzzled at the interview and barely had time to collect their thoughts or even to remember minor complaints before the examiner had assured himself of the normal function of shoulders, elbows, knees and hips and was on his way.

Contrary to popular belief about osteoarthritis, there was no association with hypertension or obesity.[14]

RHEUMATIC FEVER

The clinical features of acute rheumatic fever or acute rheumatism are too well known to require detailed review. The rheumatic feature of the disease is an acute febrile, migratory and painful arthritis lasting weeks or months, often producing complete but temporary disability and followed invariably by complete recovery. Permanent joint damage or deformity never occurs. It is essentially a disease of childhood. Its great interest from a community and public health viewpoint depends upon the regularity with which organic heart disease occurs with its attendant chronic invalidism, disability and premature death. According to insurance statistics, rheumatic fever and rheumatic heart disease are found to be the commonest causes of death between 15 and 19 years of age.[13]

The onset of rheumatic fever has long been recognized as associated with acute tonsillitis, sore throat and streptococcal infections. Thus, incidence of rheumatic fever is related to cold, damp and changeable weather, poverty, congestion, substandard living conditions and malnutrition. It has been rare in warm and sunny climates, such as Arizona and Puerto Rico, where people live outdoors, and streptococcal infections are rare.

Although rheumatic fever had long been regarded as an infectious disease, the factor of heredity also has been suspected. As information accumulated it has become clear to the author that this is a disease dependent upon 2 distinct factors, both of which must be present for the disease to develop. These are susceptibility, which depends definitely upon heredity and environment, or infection with hemolytic streptococci.

Numerous studies have been published indicating that rheumatic fever is a familial

disease, and that incidence increases as the relationship becomes more intimate with index cases; that is, siblings are affected more frequently than are parents or children, cousins or aunts, uncles and second cousins. Under certain conditions in which streptococcal infections are endemic and children live in close proximity and intimacy with each other, the disease is shown to occur in mendelian ratios of a single autosomal recessive trait. In a study published by Wilson,[26] when both parents were negative, 39 of 98 children, or 39.8 per cent were affected. When 1 parent was affected, 34 of 55 children, or 62 per cent, were affected. And in 3 families with both parents affected, all 13 children were affected.

The role of the streptococcus in the production of rheumatic fever has been summarized by Rammelkamp *et al.*[11] In studying a controlled population at Warren Air Force Base, in Wyoming, with complete medical histories, frequent physical examinations and repeated throat cultures, they found that rheumatic fever developed only after streptococcal infection, and then in only 3 per cent of patients with proved streptococcal infections. They present the data of almost identical proportions in careful studies of populations in North Carolina, Boston and Denmark. Rheumatic fever seems to appear primarily in patients who develop large amounts of antibody, as indicated by a rise in antistreptolysin titer. There seems to be no immunity to streptococcal infections, because in large family groups living in cities, nearly every individual can be seen at one time or another with streptococcal infections if he is studied closely enough for a sufficiently long period of time. Thus, it is apparent that the entire population is vulnerable to streptococcal infection, but that susceptibility to rheumatic fever is limited to the 3 per cent who develop it. The 3 per cent seem to be genetically determined on the basis of a single factor—autosomal recessive. According to this supposition, rheumatic fever attains mendelian proportions at times in dense urban populations where streptococcal infection is endemic. These proportions are not attained in environmental circumstances where streptococcal infections are rare or nonexistent, such as Puerto Rico, Arizona or Florida. Rheumatic fever is avoided or its incidence is reduced when personal hygiene is good, when streptococcal infections are not so common as in economically favored homes, when each child has his own bed and his own room, when nutrition is good and when proper treatment with antibiotics controls infection, prevents its spread and reduces the immune reaction and development of antistreptolysin titer. With continually improving housing, nutrition and prophylactic medical care, it is conceivable that rheumatic fever soon may become a completely controlled and rare disease. The same or even a larger proportion of the population will remain or become susceptible to rheumatic fever, which then may break out in a widespread epidemic, even as smallpox has done.

The 3 per cent incidence of susceptibility in the population to rheumatic fever can be used for gene frequency analysis. The findings are: 3 per cent, homozygous susceptible; 28.6 per cent, heterozygous normals who are carriers; and 68.3 per cent, homozygous normals.[18] Rheumatic fever seems to result from unusually energetic immune reaction to streptococcal infection, as shown by the antistreptolysin titer. Recognition of susceptible individuals has been attempted by observing the immune response to different innocuous antigen, but this has not been successful as yet.

When rheumatic fever is recognized, the family should be alerted to the danger of streptococcal infections, and all siblings should be treated promptly and adequately with antibiotics to cure the infection before a rise in antibodies occurs. It is conceivable that if these measures are used effectively, rheumatic fever might be eliminated.

RHEUMATOID ARTHRITIS

In a study of the genetics of rheumatoid arthritis,[23] the families of 224 patients, including 1,667 individuals, were compared with the families of 488 controls composed of 2,759 individuals. 3 per cent of all relatives of patients were found to have been affected, compared with 0.6 per cent in the control series. Of relatives over the age of 50 years, the figures obtained were 5 per cent and 0.9 per cent. Rheumatoid arthritis is found among the relatives of affected people 5 to 6 times as frequently as among others.

Among 224 families, 250 people had rheumatoid arthritis out of 1,054 brothers and sisters compared with 561 expected if the disease were inherited as a simple autosomal dominant. Gene-frequency computation, based upon 16 found to be affected among 2,759 in the control series, showed 0.58 per cent of the population picked at random with the disease. Since penetrance was about 50 per cent, the gene frequency is doubled, 0.58 to 1.16 per cent. In other words, 1.16 per cent of the general population has the proper genetic constitution for the disease, and 50 per cent of these, or 0.58 per cent, are affected. This indicates that about 12 per 1,000 are susceptible and 6 per 1,000 affected.

Many factors are present that obscure the workings of heredity and make analysis difficult or doubtful. One is the varying age of onset recorded in this series as 13 to 72 with an average age of 39.2 $\pm$ 0.9 years. Because of earlier death, one never is certain of the true number of susceptibles in the group. Another factor is the sex ratio, always higher in women than in men, with widely varying differences in proportions from series to series. Among our index cases it was 166 women to 123 men, because of nonrandom sampling and the inclusion of a large number of cases from the U. S. Veterans Hospital. Of the affected parents of index cases, 15 were mothers and 8 were fathers, a 2 to 1 ratio. Of 57 affected individuals, excluding the index cases, there were 41 women to 16 men, or a ratio of 2.5 to 1. Only 23 of 224 parents were reported to be affected as compared with 1 in each family in a dominant character. This indicates a penetrance of only 10 per cent. It is obvious that many parents suffered mild or atypical and unrecognized disease.

Although heredity plays an important part in the course of rheumatoid arthritis, other secondary conditioning factors also must be significant. They have been sought repeatedly with indifferent success. An extensive controlled study[10] was pursued in England in 1950 in which each of 532 patients with rheumatoid arthritis was paired with an unaffected person of the same age, sex and marital status. An arbitrary age of onset was assigned to each control person the same as that of his affected partner. Data were gathered and related to age of onset, real and simulated, as to tonsillitis, sinusitis, dental infections, accidents, exposure to inclement weather, faulty nutrition, unhygienic living and working conditions, pregnancy and its complications, menopause, social disaster, emotional stress and other pertinent factors. The only significant difference noted between the 2 groups seemed to be that the patients had a higher proportion of affected relatives than did the control group, and, thus, heredity was the single factor demonstrated.

Rheumatoid arthritis presents a nice theoretical problem. It is seen that gene frequency and penetrance are both low, indicating that a small proportion, 1.2 per cent, of the population is susceptible, and only one half of that number succomb to the disease. Positive secondary causative factors, such as infected teeth, tonsils and sinuses, unhygienic living and working conditions, emotional, financial and personal stress—all part of the pattern of everyday living that 99 per cent of the population sustain without developing rheumatoid arthritis—may well prove on closer examination to be critical for susceptible individuals.

Much of the evidence on the cause of

rheumatoid arthritis may have to be re-evaluated and certain current conclusions modified. The initiating factor may be as specific, as innocuous and even less closely related from the point of time to the vague, indefinite, uncertain and unrecognizable onset of rheumatoid arthritis as is a streptococcal infection in a susceptible individual to rheumatic fever. When this hypothetic initiating stimulus can affect all the population and arouse this chronic disease in less than 2 per cent of those attacked, it is small wonder that it has not been recognized as yet.

ANKYLOSING SPONDYLITIS

Ankylosing spondylitis is considered to be a definite clinical entity entirely separate from rheumatoid arthritis. That heredity plays a part in its etiology is indicated by the numerous examples of familial involvement that occur. In previous studies, about 75 instances have been cited in the literature. In our material, 7 secondary cases were found in 50 families studied.[8]

The occurrence of spondylitis in both members of 3 sets of identical twins scarcely can be explained on a basis of independent probability. In an extensive study of spondylitis in Bristol, West[24] concluded that the disease afflicted about 1 in every 2,000 of the population. Using his estimate of the incidence of the disease, the probability of 2 particular individuals being affected would be $2{,}000^2$ or 1 in 4 million. Since approximately 1 in 90 births are twins, and one third of these are identical twins, involved identical twins could be expected only once in 270 times 4 million births, or 1 in 1 billion births. Assuming spondylitis to be hereditary, it could be expected to affect both twins about once in 540,000 births. West also found that spondylitis did not follow the law of independent probabilities. In an exhaustive study of 41,907 families in Bristol, he found 74 families with 1 case of spondylitis, 8 with 2 and 3 with 3, compared with 74 families with 1, 0.065 families with 2 and 0.00003 families with 3, as expected on a basis of Poisson distribution.

In our series of 56 families there were 63 cases among 281 relatives, or an incidence of 2.5 per cent. These included 1 father, 3 brothers and 3 sisters. In a control series of 616 families with 3,335 relatives, 6 were found to be affected, or an incidence of 0.18 per cent. Thus, relatives of spondylitic patients suffer from the disease about 15 times as commonly as do members of the general population.

One striking feature of spondylitis is the sex incidence, usually given as 10 men to 1 woman. In a computation from 10 series chosen arbitrarily from the literature, the ratio proved to be 5.7 men to 1 woman. West found an incidence of 10 men to 1 woman. In our series the incidence was 55 men to 7 women. Because of this wide difference in sex incidence, penetrance had to be corrected for each sex.

Considering spondylitis as a single autosomal dominant character, in 56 affected families with 214 sibs, 62 actually were found instead of the 118 expected, indicating a penetrance of 52 per cent. Penetrance then was computed for men and women separately. It was found that 138, or 65 per cent, of 214 index cases and their sibs were men and 76, or 35 per cent, were women. Sixty-five per cent of 118 expected to be affected indicates that 77 men, and 35 per cent of 118 or 41 women, are to be expected affected. Since 55 actually are affected compared with 77 expected, penetrance is 71 per cent in men, and 7 women actually are affected compared with 41 expected affected. Penetrance is 17 per cent in women. The proportion of men to women expected to be affected is 71 to 17, or about 4 to 1. Therefore, about 80 per cent of the cases will be found in men, fairly close to the 85 per cent in the 2,321 cases cited from the literature.

Penetrance in each sex then was computed in a different way, first by dividing the series according to sex of the index case. In this computation some of the families had

to be omitted because there was a male index case where only affected women were found in the family and vice versa. For sibships containing male index cases, there are 51 families with 130 sibs and 54 affected brothers compared with 80.6 expected, showing penetrance of 67 ± 4.1 per cent. Considering 4 families with female index cases and limiting the computation to sisters alone, we find 6 of 11 sibs affected, compared with 6.5 expected, or 100 per cent penetrance. Obviously, these results are higher than those experienced in the general population, particularly in women, and are due, in part at least, to weighed sampling of the material and elimination of the families with milder involvement. This is indicated by the fact that these 2 groups include 141 sibs, instead of 214 sibs, in the 56 families. Because of this unusually high penetrance in women, correction for family of small size was applied to all families with an affected sister, and all sibs were included. The result revealed that 5 families included 19 sibs with 9 affected, compared with 10.3 ± 1.9 expected, showing 90 per cent penetrance. Bearing in mind the standard error, this may be taken as indicating perhaps complete penetrance. In sibships with a woman affected there is a stabilizing influence on the main factor, the result being a higher than ordinary penetrance. This penetrance will be discussed later. Despite the complete penetrance seen in those sibships with an affected woman, penetrance in women in the entire series was low; only 7 of the 76 women, or 9 per cent, were affected, compared with 49 of 120 men, or 41 per cent.

In the study of human heredity it now is realized more widely that the analysis of pedigrees and the conclusion drawn from the statistical tests made on the accumulated data need to be reasonably consistent with the gene frequencies in the population. It is difficult to achieve this desirable result, because many human traits commonly show a lack of penetrance of the genes involved, and there are few characteristics for which the incidence in the general population is known accurately. This also is true of spondylitis; nevertheless, it is desirable to attempt the gene-frequency analysis.

Apparently the most satisfactory data on the incidence of spondylitis are those of West,[24] who found 1 in 2,000 in the adult population of Bristol, England. He also found a ratio of 10 men to 1 woman with spondylitis. Consequently, there are 10 men and 1 woman with spondylitis in every 22,000 of the adult population.

Because of the fairly large number of reported instances of spondylitis in 2 and 3 generations, this low incidence of 1 in 2,000 of the adult population is itself evidence that spondylitis is a dominant trait. If it were a recessive, some degree of consanguinity would be expected in the 56 families. None was found.

For determining the gene frequency one needs an estimate of the penetrance, as well as the incidence, of the condition in the population. The 2 different estimates of the penetrance in men of 71 ± 3.8 per cent and 67 ± 4.3 per cent are in fair agreement, so that a round figure of 70 per cent may be taken as the penetrance in men. The estimate of the penetrance in women is somewhat less certain. From our small sampling the penetrance in women was 17 ± 4.8 per cent. But, as argued above, in these families, the penetrance is greater than in the general population. We may make another estimate of the penetrance by subtracting the 4 index cases, leaving 72 women, only 3 of whom have spondylitis, whilst on the basis of a 1 to 1 ratio 36 would be expected. This gives a penetrance of about 8 per cent in women. This agrees with West's census of the total population of Bristol; he found a ratio of 10 men to 1 woman, or a penetrance of 7 per cent. Consequently, for the calculation of gene frequency, 70 per cent penetrance in men and 10 per cent in women will be used.

Assuming the trait to be an autosomal dominant, and making the calculation first

for men with 70 per cent penetrance, 1 man in 1,540 of the population will have a genetic constitution for spondylitis. The incomplete penetrance lowers the incidence among men in the population to 1 in 2,200. With random mating of a population in genetic equilibrium, the homozygous dominants, the heterozygotes and the homozygous recessive are present in the population in the relative frequencies $d^2 + 2dr + r^2 = 1$, where d is the frequency of the dominant factor for ankylosing spondylitis and r is the frequency of the recessive normal allele. The calculation shows that for men the value of d is 0.00032, and for r the value is 0.99968; consequently, the frequency of heterozygotes $2\,dr$ is 0.00064.

If the trait is an autosomal dominant, there are as many women as men with the genetic constitution for spondylitis, so that these gene frequencies should be very similar to those obtained from the corresponding calculation from the data for women. Again, on the basis of West's figure, the incidence of women in the adult population is 1 in 22,000; but, allowing for the low penetrance, taken as 10 per cent, the frequency of women in the population with the genetic constitution becomes 1 in 2,200, so that 2,199 in every 2,200 are recessive normals. The calculation from the equation for a population in genetic equilibrium gives 0 00028 for the frequency of d, and for r the value is 0.99972; consequently, $2dr$ is 0.00056. Considering the approximate character of the incidence and the uncertainty in the value of the penetrance, these 2 values for the frequency of men and women with a heterozygous constitution for spondylitis, namely, 0.00064 and 0.00056, are in quite close agreement. In fact, rounding the figures to the 4th decimal place gives exactly the same value; that is, about 6 in 10,000 have a heterozygous genetic constitution for ankylosing spondylitis. The gene frequency d is so low that for the present discussion the probable incidence of homozygous dominants d^2 in the population may be disregarded.

The most plausible, nevertheless still tentative, conclusion from the analysis of the data is that ankylosing spondylitis is due to an autosomal dominant factor that has about 70 per cent penetrance in males and perhaps about 10 per cent or slightly greater penetrance in females. This conclusion is quite consistent with the analysis by inspection of significant pedigrees, with a numerical test on pooled data and with the estimated gene frequency in the population.

In regard to the difference in penetrance between males and females, there is a further peculiarity that is interesting in itself and may become of considerable importance in the further study of human genetics, especially if it should prove to be of common occurrence. It seems that in sibships containing an affected woman there is a stabilizing effect of the main factor, and penetrance is higher than the average in such families. Penetrance was found to be nearly complete in the 5 sibships containing an affected female in the present series. This seemed to be of sufficient interest to warrant further investigation. Further pedigrees, including women affected with spondylitis, were sought in the literature and in personal communication. Care was taken to accept only family histories in which all the sibs, normal as well as affected, were described. 15 families were assembled, including 77 sibs, of whom 37 were reported to be affected. After correction for families of small size 40.6 ± 4 were expected to be affected. These families showed complete penetrance.

GOUT

Gout long has been recognized as a genetic disease, but heredity has been irregular, and no definite pattern of heredity has been recognized. Since gout is associated with hyperuricemia, and hyperuricemia has been found to precede clinical gout for many years, the study of the heredity of gout has become the study of hereditary or idiopathic

hyperuricemia. More reliable results were obtained by using serum uric acid determination instead of whole blood.

In a family study of the disease,[22] including 52 index cases, it was found that 2 of 11 mothers, 4 of 23 brothers, 5 of 25 sisters and 5 of 33 sons had hyperuricemia, although every one of 45 daughters was normal. In 16 families with an affected father, only 3 of 27 sons were involved. In 5 families with a mother involved, 6 of 11 sons were hyperuricemic.

Involvement of women with hyperuricemia was rare but significant. Of 4 sibships with involved women, a total of 6 were examined and found to be involved. Of 4 brothers of these women, 3 were involved. 9 of the 10 people in these sibships involving affected women had hyperuricemia.

Study of the families of this series justifies the conclusion that the genetic peculiarities of hyperuricemia are mainly the expression of an autosomal dominant gene that lacks penetrance in both sexes but with much lower penetrance in the female than in the male, or 14 per cent in females, 84 per cent in males.

The data are consistent with the view that gouty arthritis may develop in an individual of either sex who possesses sufficiently elevated serum urate level for sufficient time, this being less probable in heterozygous females owing to a lower normal level in women and to a lessened effect of the pathologic gene in this sex. It is shown that the gene for essential hyperuricemia must be considerably more common than one would suspect from the incidence of clinical gout, and, therefore, homozygotes should be observed occasionally for this gene Plasma urate levels have been found to be higher in men than in women. This is true of normal people, of gouty patients and of hyperuricemic relatives of gouty patients. Often, men who inherited hyperuricemia do not show it until after puberty. Usually, women do not show it until after the menopause. There is some correlation between the duration and the magnitude of hyperuricemia and the onset of gout. These factors all help to explain why clinical gout is so much more common in men than in women and why the incidence increases with age.

Recently Wolfson[27] advanced a new theory of gout. He believes that the inherited susceptibility to gout depends upon an abnormal male sex hormone from the adrenal that interferes with uric acid excretion by the kidney and thereby produces hyperuricemia. These abnormal male sex hormones produce effects abnormally marked on uric acid metabolism but normally on masculinity. Attacks of clinical gout depend upon a second endocrine disturbance, an inability to produce promptly and in adequate amounts needed 11-oxysteroids during periods of stress due to failure of the pituitary to release adequate amounts of ACTH. This explains why ACTH is effective in relieving acute attacks of gouty arthritis.

OSTEOARTHRITIS OF THE HIP

The problem of heredity in joint diseases of the hip is an extremely complicated one. This depends upon the fact that osteoarthritis of this joint may arise from a variety of causes and have varying etiologies. Unfortunately, in the advanced, or well-developed, state of osteoarthritis it is difficult or impossible to recognize these preceding predisposing factors. The causes of osteoarthritis of the hip include previous fracture, traumatic dislocation and aseptic bone necrosis, all due to injury without relation to genetics. Congenital dysplasia of the hip, coxa plana, slipped epiphyses and Perthes' disease also are followed occasionally by arthritis. In each of these diseases heredity has been suspected or demonstrated. Several of these conditions may have the same fundamental hereditary defect, although they differ in their manifestations as to age of onset, sex distribution, proportion of unilateral and bilateral involvement. Since these conditions are not all apparent at birth, are diagnosed with uncertainty, and only then

after careful roentgenographic investigation, and since some of them tend to heal spontaneously, family pedigrees show irregular involvement with wide deviation from expected mendelian proportions. Furthermore, no convincing figures have been assembled to show what proportion of patients with these primary genetic characters develop osteoarthritis, nor does anyone know what proportion of osteoarthritis of the hip depends upon such genetic characters.

Graver-Duvernay[5] states unequivocably that arthritis develops only on pre-existing deformities of the hip, usually subluxation, coxa plana or coxa vara. That this is not entirely true is shown by the author's recent observation of well-developed osteoarthritis of 1 hip in a 60-year-old woman who brought with her a roentgenogram of the pelvis, taken 5 years earlier, that gave absolutely no sign of pathologic involvement. There was no recognizable injury or infection in the interval. Graver-Duvernay quotes and agrees with Bezaçon, M. P. Weil, Calot and Munk-Jansen, who claim that 50 per cent of cases of osteoarthritis of the hip are due to previous congenital deformity.

The outstanding number of hip deformities in man has been attributed to the large brain and the wide pelvis of this species.[9] The well-known sex difference in size of the pelvis accounts for the greater frequency of hip disease in the female. That various anomalies are related is shown by the fact that several occur in the same person such as coxa vara and coxa plana, coxa plana and coxa valga, coxa valga and slipped epiphysis, coxa valga and malum coxae. These defects often alternated in various members of the same family. They also alternate with subluxation and congenital hip dysplasia. These lesions have a regular progression regarding age of occurrence, that of coxa vara being from 1 to 3 years, coxa plana from 3 to 10 years, coxa valga from 7 to 13 years, and malum coxae 14 years and over. The defective hip joint results from altered stresses in the head and the neck in a flattened hip socket.

The dysplastic hip is underdeveloped and tends toward the infantile form.[1] Its form is not round but egg shaped; its position is a little farther front and lateral. The neck and the head in intra-uterine life stand in sagittal position. The fetal form of the pelvis begins to change at the end of pregnancy. The socket and the head travel backward and laterally to the normal position of the adult. If the fetal form remains in extra-uterine life, we have a defective hip joint that does not grow and becomes a congenital hip luxation or coxa plana or the like. The heredity of this lesion and the analogy of the human embryonal form of the hip to that of the primates, particularly the gorilla, bespeak a failure of development.

Wiberg[27] selected 50 men and 50 women between the ages of 20 and 35 with normal hips and re-examined them later in life. He found that 19 of these normal hips had developed osteoarthritis, as proved roentgenographically. He also saw 5 of 19 cases of maldeveloped hips that healed spontaneously and 3 hips normal in childhood that developed poor acetabula in later life. Sclerosis of the roof of the acetabula appeared more often in defective than in normal hips.

Considerable evidence is available that supports the hereditary nature of diseases of the hip and the relationship of luxation, subluxation, dysplasia and Legg-Calvé-Perthes' disease. Francillon[3] quotes a pedigree from Lange which, in 3 generations, included 2 cases of congenital hip dislocation and 17 other cases of coxa vara, subluxation and arthrosis, coxa deformans or osteoarthritis. In the last, the osteoarthritis occurred in cases of subluxations that had been completely without symptoms or nearly so. Isigkeit noted a hereditary influence in 20.2 per cent of his cases, Scaglietti in 22.7 per cent, but Francillon, depending entirely upon written questionnaires by mail, found only 6.1 per cent. Isigkeit called the character a recessive one; von Verschuer and

Lange, an irregular dominant. It is well known that incidence varies considerably in different races and different geographic localities.

Hass[6] names at least 10 authors who have presented pedigrees of hereditary transmission of congenital hip dislocation. He says that Vogel noted hereditary effects in 30 per cent of 200 cases, though direct transmission from parent to child was found only 6 times. Poli noted a hereditary influence in 26.32 per cent of 8,610 cases. In Japan, Hayashi and Matsuoka found it in 14.7 per cent of 1,096 cases, though familial involvement was recognized in only 3.4 per cent. Faber found that the latent form of the hereditary anlage is at least as frequent as the disease itself, so that even so-called isolated cases can be hereditary. In 19 isolated cases that were studied carefully by roentgenograms, other members of the families invariably were found with various stages from unrecognized subluxation to actual dislocation. The most probable explanation of the hereditary manifestation seen is that of van Hooff, that congenital hip dysplasia is inherited as an irregular dominant.

Zinsli[28] has investigated 43 members in 3 generations of 1 family and found 14 of them suffering from hip and 5 from knee disturbances. These findings depend upon a hereditary defect that is dominant. The primary disease is a deformation of the epiphysis leading to secondary arthrotic changes. It affects men primarily, but definitely it is not sex linked. It is a congenital dysplasia with luxation

In a recent monograph on Legg-Calvé-Perthes disease, Goff[4] states that this constitutes about 15 to 17 per cent of all hip disease. He states that heredity can be demonstrated in about 20 per cent of cases, and that it seems to depend upon a recessive with varying penetrance. There is no evidence of sex linkage or sex influence, yet there is definite linkage because of [illegible] y in sex predominance, body bu[illegible] incidence and bilaterality and [illegible] traits These manifest themse[illegible]

retardation, particularly of the hip. The American Indian and the Negro do not seem to contact the disease.

Rüther[12] has assembled the evidence of familial involvement of slipped epiphyses. These include 1 instance in twin sisters, 1 of involvement of 3 of 6 siblings, 1 of 2 brothers in a family of 12 children and 2 children in a family of 14. There also were a father and son. Rüther describes a mother and 2 daughters affected.

At least one attempt has been made to distinguish osteoarthritis of the hip in the final stage based on dysplasia or underdevelopment and osteoarthritis of a formerly normal hip. The difference depends upon the site of the lesion. If decrease in joint space and condensation of joint surfaces occur at the top of the head of the femur, it indicates that osteoarthritis is developing in a normally developed hip. If there is increased distance between the medial surface of the head and the floor of the acetabulum, the medial type is present; the head is being forced out of the socket, and this indicates disease in an underdeveloped hip. The author Hermodsson[7] admits that this distinction is difficult or impossible in advanced disease, and that it is advisable to make decision early in its development.

There are wide racial and geographic differences in congenital dysplasia of the hip and, consequently, of osteoarthritis of this joint. In certain areas of Switzerland, as well as of Brittany, the incidence of congenital dysplasia is high; the descendants of immigrants who settled there centuries ago have remained separate and isolated from their neighbors, with a high incidence of inbreeding and certain anatomic characteristics. In both areas congenital dysplasia is associated with short stature and broad heads. In Switzerland, [illegible] is a high incidence of red hair.[3] Des[illegible] described these people in Br[illegible] [illegible]ficulties, has suggested that the [illegible] or method of prevention [illegible] is to advise marriage with [illegible]

CONCLUSIONS

This discussion has been limited almost entirely to a presentation of the results of various investigations that have been pursued in the past 15 years. Details about the literature on the subject, the methods of investigation, the actual collection of the data, the computation of the statistics and a discussion of the conclusions will be found in the literature cited.

At first glance it may appear that the study of heredity in joint disease is largely an academic subject without practical significance. The study of Heberden's nodes was undertaken first because the disease was clear cut and the diagnosis reliable. Two types of Heberden's nodes were distinguished, but, more important, it was demonstrated that Heberdens' nodes were an osteoarthritis of the finger joints alone and not an indication of a generalized or a crippling disease. Because of the close association of this disease with the menopause, it is assumed that it might be prevented, or at least delayed, if the hormonal or the chemical effects of the menopause could be avoided or counteracted.

Exact knowledge of the heredity of rheumatic fever has emphasized the importance of prophylactic measures against streptococcal infections in those who are susceptible. It is possible that a similar procedure may be developed in the future for the prophylaxis of rheumatoid arthritis.

The knowledge that ankylosing spondylitis runs in families has prompted some alert rheumatologists to investigate the brothers of patients by roentgenographic examination and the early identification of additional cases. By investigating the brothers of gouty patients, those susceptible have been identified. This has the distinct advantage of ensuring prompt and accurate diagnosis of gout when an acute arthritis occurs.

It is hoped that accurate diagnosis and prompt treatment of dysplasia of the hip will reduce the incidence of osteoarthritis of the hip in middle or late life.

While great progress has been made already in the field of human genetics, it is hoped that continued attention to this type of investigation will lead to further exact knowledge about different diseases, with resultant benefit to the patient.

REFERENCES

1. Böhn, M.: Entstehung der angeborenen Hüftverrenkung, Ztschr. orthop. Chir. 55: 566, 1931.
2. Desse, G.: Sur les luxations de hanches, coxarthroses et coxites; étude génétique, Rev. rhum. 21:161, 1954.
3. Francillon, M. R.: Beitrag zur Kenntnis der angeborenen Hüftgelenk-verrenkung, Stuttgart, Enke, 1937.
4. Goff, C. W.: Legg-Calvé-Perthes' Syndrome, Springfield, Ill., Thomas, 1954.
5. Graver-Duvernay, J.: L'aspect clinique des arthrites chroniques de la hanche d'origine congénital, Rev. rhum. 5:304, 1938.
6. Hass, J.: Congenital Dislocation of the Hip, Springfield, Ill., Thomas, 1951.
7. Hermodsson, I.: On the roentgenologic appearance of osteoarthrosis deformans in the hip joint, acta radiol., Supp. 66, 1947.
8. Hersh, A. H., Stecher, R. M., Solomon, W. M., Wolpaw, R., and Hauser, H.: Heredity in ankylosing spondylitis; a study of fifty families, Am. J. Human Genet. 2:391, 1950.
9. Jansen, M.: Large brain, wide pelvic girdle and the outstanding number of hip anomalies in man: coxa vara, coxa fracta, coxa plana, coxa valga, slipping epiphysis, malum coxa, J. Bone & Joint Surg. 11: 461, 1929.
10. Lewis-Faning, E.: Report on an enquiry into the aetiological factors associated with rheumatoid arthritis, Ann. Rheumat. Dis., Supp. 9, 1950.
11. Rammelkamp, C. H., Wannamaker, L. W., and Denny, F. W.: The epidemiology and prevention of rheumatic fever, Bull. New York Acad. Med. 28:321, 1952.
12. Rüther, H.: Ursachen und Behandlung der jugendlichen Hüftkopflösung, Stuttgart, Enke, 1954
13. Statistical Bulletin: Metropolitan Life Insurance Co. 33:8-11, 1952.
14. Stecher, R. M.: Heberden's nodes: the association of hypertension and obesity to degenerative joint disease of the fingers, J. Lab. & Clin. Med. 31:687, 1946.

Lange, an irregular dominant. It is well known that incidence varies considerably in different races and different geographic localities.

Hass[6] names at least 10 authors who have presented pedigrees of hereditary transmission of congenital hip dislocation. He says that Vogel noted hereditary effects in 30 per cent of 200 cases, though direct transmission from parent to child was found only 6 times. Poli noted a hereditary influence in 26.32 per cent of 8,610 cases. In Japan, Hayashi and Matsuoka found it in 14.7 per cent of 1,096 cases, though familial involvement was recognized in only 3.4 per cent. Faber found that the latent form of the hereditary anlage is at least as frequent as the disease itself, so that even so-called isolated cases can be hereditary. In 19 isolated cases that were studied carefully by roentgenograms, other members of the families invariably were found with various stages from unrecognized subluxation to actual dislocation. The most probable explanation of the hereditary manifestation seen is that of van Hooff, that congenital hip dysplasia is inherited as an irregular dominant.

Zinsli[28] has investigated 43 members in 3 generations of 1 family and found 14 of them suffering from hip and 5 from knee disturbances These findings depend upon a hereditary defect that is dominant. The primary disease is a deformation of the epiphysis leading to secondary arthrotic changes. It affects men primarily, but definitely it is not sex linked. It is a congenital dysplasia with luxation

In a recent monograph on Legg-Calvé-Perthes disease, Goff[4] states that this constitutes about 15 to 17 per cent of all hip disease. He states that heredity can be demonstrated in about 20 per cent of cases, and that it seems to depend upon a recessive with varying penetrance. There is no evidence of sex linkage or sex influence, yet there is definite linkage because of similarity in sex predominance, body build, age of incidence and bilaterality and other complex traits These manifest themselves in growth retardation, particularly of the hip. The American Indian and the Negro do not seem to contact the disease.

Rüther[12] has assembled the evidence of familial involvement of slipped epiphyses. These include 1 instance in twin sisters, 1 of involvement of 3 of 6 siblings, 1 of 2 brothers in a family of 12 children and 2 children in a family of 14. There also were a father and son. Rüther describes a mother and 2 daughters affected.

At least one attempt has been made to distinguish osteoarthritis of the hip in the final stage based on dysplasia or underdevelopment and osteoarthritis of a formerly normal hip. The difference depends upon the site of the lesion. If decrease in joint space and condensation of joint surfaces occur at the top of the head of the femur, it indicates that osteoarthritis is developing in a normally developed hip. If there is increased distance between the medial surface of the head and the floor of the acetabulum, the medial type is present; the head is being forced out of the socket, and this indicates disease in an underdeveloped hip. The author Hermodsson[7] admits that this distinction is difficult or impossible in advanced disease, and that it is advisable to make decision early in its development.

There are wide racial and geographic differences in congenital dysplasia of the hip and, consequently, of osteoarthritis of this joint. In certain areas of Switzerland, as well as of Brittany, the incidence of congenital dysplasia is high; the descendants of immigrants who settled there centuries ago have remained separate and isolated from their neighbors, with a high incidence of inbreeding and certain anatomic characteristics. In both areas congenital dysplasia is associated with short stature and broad heads. In Switzerland, there also is a high incidence of red hair.[3] Desse,[2] who has described these people in Brittany and their difficulties, has suggested that the best treatment or method of prevention of hip deformities is to advise marriage with people of different constitutions.

CONCLUSIONS

This discussion has been limited almost entirely to a presentation of the results of various investigations that have been pursued in the past 15 years. Details about the literature on the subject, the methods of investigation, the actual collection of the data, the computation of the statistics and a discussion of the conclusions will be found in the literature cited.

At first glance it may appear that the study of heredity in joint disease is largely an academic subject without practical significance. The study of Heberden's nodes was undertaken first because the disease was clear cut and the diagnosis reliable. Two types of Heberden's nodes were distinguished, but, more important, it was demonstrated that Heberdens' nodes were an osteoarthritis of the finger joints alone and not an indication of a generalized or a crippling disease. Because of the close association of this disease with the menopause, it is assumed that it might be prevented, or at least delayed, if the hormonal or the chemical effects of the menopause could be avoided or counteracted.

Exact knowledge of the heredity of rheumatic fever has emphasized the importance of prophylactic measures against streptococcal infections in those who are susceptible. It is possible that a similar procedure may be developed in the future for the prophylaxis of rheumatoid arthritis.

The knowledge that ankylosing spondylitis runs in families has prompted some alert rheumatologists to investigate the brothers of patients by roentgenographic examination and the early identification of additional cases By investigating the brothers of gouty patients, those susceptible have been identified. This has the distinct advantage of ensuring prompt and accurate diagnosis of gout when an acute arthritis occurs.

It is hoped that accurate diagnosis and prompt treatment of dysplasia of the hip will reduce the incidence of osteoarthritis of the hip in middle or late life.

While great progress has been made already in the field of human genetics, it is hoped that continued attention to this type of investigation will lead to further exact knowledge about different diseases, with resultant benefit to the patient.

REFERENCES

1. Böhn, M.: Entstehung der angeborenen Hüftverrenkung. Ztschr. orthop. Chir. 55: 566, 1931.
2. Desse, G.: Sur les luxations de hanches, coxarthroses et coxites; étude génétique, Rev. rhum. 21:161, 1954.
3. Francillon, M. R.: Beitrag zur Kenntnis der angeborenen Hüftgelenk-verrenkung, Stuttgart, Enke, 1937.
4. Goff, C. W.: Legg-Calvé-Perthes' Syndrome, Springfield, Ill., Thomas, 1954.
5. Graver-Duvernay, J.: L'aspect clinique des arthrites chroniques de la hanche d'origine congénital, Rev. rhum. 5:304, 1938.
6. Hass, J.: Congenital Dislocation of the Hip, Springfield, Ill., Thomas, 1951.
7. Hermodsson, I.: On the roentgenologic appearance of osteoarthrosis deformans in the hip joint, acta radiol., Supp. 66, 1947.
8. Hersh, A. H., Stecher, R. M., Solomon, W. M., Wolpaw, R., and Hauser, H.: Heredity in ankylosing spondylitis; a study of fifty families, Am. J. Human Genet. 2:391, 1950.
9. Jansen, M.: Large brain, wide pelvic girdle and the outstanding number of hip anomalies in man: coxa vara, coxa fracta, coxa plana, coxa valga, slipping epiphysis, malum coxa, J. Bone & Joint Surg. 11: 461, 1929.
10. Lewis-Faning, E.: Report on an enquiry into the aetiological factors associated with rheumatoid arthritis, Ann. Rheumat. Dis., Supp. 9, 1950.
11. Rammelkamp, C. H., Wannamaker, L. W., and Denny, F. W.: The epidemiology and prevention of rheumatic fever, Bull. New York Acad. Med. 28:321, 1952.
12. Rüther, H.: Ursachen und Behandlung der jugendlichen Hüftkopflösung, Stuttgart, Enke, 1954.
13. Statistical Bulletin: Metropolitan Life Insurance Co. 33:8-11, 1952.
14. Stecher, R. M.: Heberden's nodes: the association of hypertension and obesity to degenerative joint disease of the fingers, J. Lab. & Clin. Med. 31:687, 1946.

15. ———: Heberden's nodes: heredity in hypertrophic arthritis of the finger joints, Am. J. M. Sc. 201:801, 1941.
16. ———: Heberden's nodes: the incidence of hypertrophic arthritis of the fingers, New England J. Med. 222:300, 1940.
17. ———: Heberden's nodes: their relation to other degenerative joint diseases, Arch. Phys. Med. 27:409, 1946.
18. ———: Heredity in Joint Diseases, Documenta Rheumatologica Geigy. To be published.
19. Stecher, R. M., Beard, E. E., and Hersh, A. H.: Heberden's nodes: the relationship of the menopause to degenerative joint disease of the fingers, J. Lab. & Clin. Med. 34:1193, 1949.
20. Stecher, R. M., and Hersh, A. H.: Heberden's nodes: the mechanism of inheritance in hypertrophic arthritis of the fingers, J. Clin. Invest. 23:699, 1944.
21. Stecher, R. M , Hersh, A. H., and Hauser, H.: Heberden's nodes: the family history and radiographic appearance of a large family, Am. J. Human Genet. 5:46, 1953.
22. Stecher, R. M., Hersh, A. H., and Solomon, W. M.: The heredity of gout and its relationship to familial hyperuricemia, Ann. Int. Med. 31:595, 1949.
23. Stecher, R. M., Hersh, A. H., Solomon, W. M., and Wolpaw, R.: The genetics of rheumatoid arthritis; analysis of 224 families, Am. J. Human Genet. 5:118, 1953.
24. West, H. F.: A Study of Ankylosing Spondylitis (Thesis), Bristol, England, 1948.
25. Wiberg, G., Studies on dysplastic acetabula and congenital subluxation of the hip joint, with special reference to the complication of osteoarthritis, Acta chir. scandinav., Supp. 58, 1939.
26. Wilson, M. G.: Rheumatic Fever, New York, Commonwealth Fund, 1940.
27. Wolfson, W. Q., Guterman, H. S., Levine, R., Cohn, C., Hunt, H. D., and Rosenberg, E. F.: An endocrine finding apparently characteristic of gout: very low urinary 17-ketosteroid excretion with clinically normal androgenic function, J. Clin. Endocrinol. 9:497, 1949.
28. Zinsli, P. E.: Arthrosis deformans der Hüfte als Erbleiden (Epiphysen-Deformation) (Inaugural Dissertation), Zurich, Art. Institut Orell-Füssli A. G., 1946.

Genetica del Morbos Articular

Summario in Interlingua

Es discutite le genetica de plure differente morbos articular. A causa de lor differentias quanto a etate del patiente al tempore del declaration del morbo, a distribution sexual, al articulationes afficite per illos, al curso clinic, e al prognosis, illos debe esser considerate como entitates distincte que require discussiones separate.

Le nodos de Heberden o osteoarthritis del articulationes digital es sexualmente influentiate: dominante in feminas e recessive in homines. Le declaration es apparentemente relationate al menopause, e le penetration es complete in le octave decennio del vita. Le frequentia genic monstra affection homozygotic de 3 pro cento e heterozygotic de 27 pro cento.

Febre rheumatic resulta de infection streptococcal in individuos susceptible. Le susceptibilitate es hereditabile como tracta recessive mendelian; circa 3 pro cento del population es afficite. Le morbo mesme resulta de un intense reaction immunologic.

Arthritis rheumatoide es apparentemente hereditabile como tracto autosomal a factor unic; 1,2 pro cento del population es geneticamente susceptibile, e un medietate de iste procentage es afficite.

Spondylitis ankylopoietic es apparentemente hereditabile como tracto autosomal unic con frequentia genic de susceptibilitate amountante a 6 per dece milles in le population total e penetration in 70 pro cento del homines e 10 pro cento del feminas. In fraternos de feminas afficite, le penetration es afficite de maniera a attinger 100 pro cento in homines e feminas.

Le gutta depende de un hyperuricemia que es hereditabile como gen dominante

autosomal con penetration de 84 pro cento in homines e 14 pro cento in feminas. Idiopathic hyperuricemia depende de un anormal hormon sexual que disturba le excretion de acido uric. Attaccos de gutta clinic depende de un anormalitate additional, i.e. le incapacitate de producer promptemente e in quantitates adequate le 11-oxysteroides requirite in periodos de stress. Le ration es que le glandula pituitari non emitte adequate quantitates del droga.

Osteoarthritis del coxa se disveloppa usualmente in un articulation ledite o subdisveloppate. Multes de iste imperfectiones depende de conditions que es geneticamente determinate. Tal conditiones es dysplasia congenite, morbo de Legg-Calvé-Perthes, coxas plan, e epiphyses dislocate.

5

Estimation of Mutation Rates in Man

J. N. Spuhler, Ph.D.*

Each of the several thousand genes existing today in a human individual probably has remained constant in its biophysical make-up for several thousands of years. Direct empiric evidence in support of this statement is of course rare. Perhaps the longest documented period of stability for a known human gene is the case of transmission through 14 generations (starting A.D. 1390) of a dominant gene for symphalangism of the first and the second fingers in an English kindred described by Drinkwater (1917). Analogy from experimental animals and plants as well as genetic theory supports an assumption of the general stability of genes. As the normal human organism grows and reproduces through processes of growth, division and fusion of cells, each gene must, in general, reproduce repeatedly its own fine structure in an exact manner. Sometimes, however, a daughter gene is not an exact duplicate of its mother gene and as a result of this "mutation" a new type of gene comes into being. Such new mutant genes have a different effect on the development of the organism, and they are capable of reproducing their own new fine structure. The combined properties of stability (exact self-reduplication) and mutability (self-reduplication after the new type) of genes make possible the existence of hereditary variability.

In this paper the author has a triple purpose. (1) to give a brief account of mutation and of certain methods for estimation of mutation rates in man, (2) to give a tabular summary of some of the available estimates, and (3) to mention the prospects for future research on mutation and mutation rates in man, using material of interest to orthopaedic surgery and others interested in inherited variations of the bones and the joints.

Mutation refers to the appearance of a trait, subsequently inherited, in an individual whose ancestors did not possess the trait. The first phenotypical appearance of the trait may not follow immediately upon the origin by mutation of the new allele. If the new allele is an autosomal or sex-linked dominant with full penetrance, then the first phenotypical expression of the gene will be in the generation immediately following the event of mutation. If the new allele is an autosomal recessive, the first phenotypical expression of the gene may be separated by several generations from the mutation event, the gene being carried in a heterozygous state. If the new allele is a sex-linked recessive it may be carried without phenotypical expression for several generations in heterozygous females but would be expressed phenotypically at once in hemizygous males.

For the purposes of this paper we shall consider only the macroscopic, phenotypical results of mutation and not the submicroscopic, physiochemical events which happen when a gene mutates. Actually, the latter are not established in detail (see, for example, Goldschmidt 1955). However, enough is known on the phenotypical level to allow

* Department of Human Genetics, University of Michigan Medical School.

some insight into mutation as a factor in disease and certain malformations (see Neel & Schull 1954, Sorsby 1953).

Mutation may be regarded as a change in pattern of the atoms, the electrons and the energy-quanta in or near the gene in question. These events are of sufficiently small scale so that one gene in a cell may mutate while the thousands of other genes in the same cell remain unchanged. Mutation may be induced by ionizing radiation which reaches the gonads. All types of ionizing radiation are effective—x-rays, alpha, beta, gamma and cosmic rays, neutrons, etc.—whether from sources internal or external to the body. Other mutagenic agents, for example temperature and chemical shocks, are known. Mutation tends to change genes in a limited number of ways, some of which are recurrent, and this allows us to speak of the rate of mutation of a given gene. When a given gene mutates in a number of different ways multiple alleles are formed (for example, the 3 alleles for the 4 classical ABO blood groups).

Mutations may occur in embryonic or adult stages and in germinal or somatic tissues, but, of course, only germinal mutations are inherited. Fundamental (e.g., agenesis of the notochord) or superficial (extra hair on the ears) changes may be produced. In general, the greater the change the greater the disadvantage to the organism in a standard environment and—in the absence of therapy—the less the survival value in that environment. Cases are known where highly similar mutations occur in what somewhat loosely may be termed homologous genes of related species. For example, clinical hemophilia determined by a gene located in the nonhomologous part of the X-chromosome is known in man, dogs and horses. Achondroplasia (chondrodystrophy) is reported for man, Dexter cattle, chickens and rabbits. Also, different nonhomologous genes may mutate to produce highly similar phenotypical effects (compare, for example, hemophilia and Christmas disease in man).

In fruit flies (*Drosophila* sp.) the effect of mutation produced most frequently is the semilethal and the deleterious, the ratio of *visibles : lethals : semilethals and deleterious* being about 1:15:20. Mutant genes act to kill the organism, often in early stages in the life cycle. When not lethal, mutations are often deleterious, the action of the mutant gene tending to harm rather than to help organic function in the standard range of environments. In any highly complex entity like a gene (whose lineage must be supposed to have survived a long series of recurrent mutations in the larger gene pool), the vast majority of random changes in fine structure would be expected to have a low survival value. Beneficial mutations should be expected to occur very rarely. A large proportion of new gene mutations do not survive to the next generation. Most (but not all, as we shall see below) mutant genes are recessive to their parent allele; such recessive genes are not manifest in the phenotype unless gametes from two individuals with the same (in a functional sense) mutant genes fuse to form a homozygote. Thus many mutant genes are lost from the population before they are shown phenotypically.

In human breeding populations there are 4 processes that change the frequency of genes: mutation; gene flow, or introduction from outside through migration; selection; and genetic drift. Taken together, these 4 processes are so defined that they give an exhaustive list of the immediate, systemic modes of change in gene frequency. In what follows we shall be concerned with so-called "point mutations" of genes and not with "mutations" which may result from grosser sorts of chromosomal aberration. A *mutant* is an individual carrying a mutated gene. Mutation *rate* will refer to the rate of mutation per gene per generation.

In a number of ways mutation and selection are conceptually kindred processes. We sometimes use knowledge about selection rates to infer knowledge about mutation

The equilibrium condition is

$$u = c\ (1 - f)x$$

Following a point of view reported by Krooth (1953), we may illustrate the calculations by considering the mutation rate for achondroplasia, an autosomal dominant inherited chondrodystrophy. Achondroplasia is sufficiently rare that we can assume that x is small; that all or nearly all cases are heterozygotes; and that we can ignore matings of abnormal with abnormal. Assume there are N normal individuals in the population and a small fraction of these, B, marry achondroplasics. If the normals have on the average b offspring in the next generation, then the number of children in the next generation from normal $\times$ normal matings will be $\frac{1}{2}(N - B)b$ of whom $u(N - B)b$ will be achondroplasics and $(1 - 2u)\frac{1}{2}(N - B)b$ will be normal. There are approximately Nx achondroplasics in the population and these are parents of $Nxbf$ children (as also are the B normals who marry achondroplasics) of which $Nxbf/2$ will be achondroplasic and $Nxbf/2$ will be normal. Thus if the population is in equilibrium the frequency of achondroplasia in the next generation will be:

$$\frac{Nxbf/2 + u(N - B)b}{Nxbf + \frac{1}{2}(N - B)b} \Big/ \frac{\frac{1}{2}xf + u(1 - B/N)}{xf + \frac{1}{2}\ (1 - B/N)} = x$$

On the assumption that x is small, that matings between abnormals are very rare, that there are Nx achondroplasics, that $f < 1$, that u is small, that B/N is of the same order as x, we can approximate the above formula by

$$(\tfrac{1}{2}\ xf + u)/\tfrac{1}{2} = x$$

or, in terms of mutation rate for an autosomal dominant ($c = \frac{1}{2}$),

$$u = \tfrac{1}{2}(1 - f)x$$

Values of c for certain other modes of inheritance together with formulas for u are:

Sex linked recessive,

$$c = \tfrac{1}{3};\ u = \tfrac{1}{3}(1 - f)x$$

Sex linked dominant,

$$c = \tfrac{2}{3};\ u = \tfrac{2}{3}(1 - f)x$$

Holandric (Y-borne),

$$c = 1;\ u = (1 - f)x$$

A classic genetic study by Mørch (1941) of achondroplasic dwarfs is perhaps the best known—but not the most accurate—material on mutation rates in man (see Haldane 1948, Neel 1952, Popham 1953, Krooth 1953, Neel and Schull 1954, Nachtsheim 1954, and Vogel 1954a). The sample collected by Mørch in Denmark included 10 achondroplasic babies born out of a total of 94,075 births over a 30-year period in the Rigshospital in Copenhagen. Although infants with this condition are subject to high mortality rates, the adult dwarfs are often vigorous and sometimes reproduce. The total Danish sample of 108 achondroplasics recorded by Mørch had a total of 27 offspring. Inspection of pedigrees shows that the cases under consideration were inherited as an autosomal dominant with high penetrance and rather constant expression. Stevens reported a similar mode of inheritance for this condition in an extensive family study made in Utah; however, a few families showing recessive modes of inheritance have been described (Falls 1953). Sporadic cases where both parents and all known ancestors have normal skeletal development are relatively common compared with familial cases where usually only one parent of the dwarf child is achondroplasic.

In order to estimate the mutation rate for an autosomal dominant condition like achondroplasia by the direct method we first need to obtain an incidence estimate, s/t, the ratio of *sporadic* (i.e., nonfamilial cases, by hypothesis newly mutant) cases to total cases. 8 of the 10 cases in the 30-year Rigshospital sample were sporadic. Thus by the direct method the mutation rate is

$$u = \tfrac{1}{2}(8/94{,}073) = 0.0000425,\ \text{or},\ 4.25 \times 10^{-5}$$

In the indirect method we use the total (sporadic plus familial) incidence which is 10/94,075 in the hospital sample. The rela-

tive reproductive fitness, f, of achondroplasics may be obtained from Mørch's data for the larger Danish sample. Since the 108 dwarfs had 27 children while their 457 normal siblings had a total of 582 children,

$$f = (27/108)/(582/457) = 0.1963$$

(see below for a bias inherent in this method of estimating f) and by the indirect method the mutation rate is

$$u = \tfrac{1}{2}(1 - 0.1963)\ (10/94{,}075) = 4.27 \times 10^{-5}$$

It should be noted that the incidence figure used in the direct method (8/94,073) is very close to that used in the indirect method (10/94,075) which accounts in part for the good agreement in the rate estimates obtained from the two methods.

Before considering some of the assumptions presupposed by use of the methods outlined above, attention should be called to some other methods for estimation of mutation rates in man which cannot be reviewed here. Neel and his associates (1949) have used data on the frequency of recessive traits in populations where the mean population coefficient of inbreeding is known with some accuracy to estimate the rate of mutation to recessive genes. Dahlberg (1948) suggested a modification of the persistence method of Danforth based on ascertainment of the number of ancestors manifesting a given trait Lenz (1952) suggested a modification of the direct method which (if suitable data were available) could provide estimates of the mutation rate to sex-linked recessive lethals in man.

Use of both methods outlined here assumes that the phenotypical variation observed can be attributed to simple, known modes of inheritance where the genes in question occupy a single chromosomal locus. If several loci are involved in the production in what is taken to be a single clinical or phenotypical entity, the estimated rate will be too high Unfortunately, unless information is available on crosses between the different family lines or populations of interest in a particular investigation we have no good criterion to establish genetic homogeneity (a difficulty, for example, in genetic studies of progressive muscular dystrophy). Also, it is often difficult to distinguish sporadic cases due to environmental factors (phenocopies) from those due to gene mutations, especially in dominant characters with strong negative selection values (for example, in achondroplasia, or in retinoblastoma). And, in diseases like retinoblastoma which often occur unilaterally, it is difficult (unless there are offspring) to distinguish germinal mutation with lack of manifestation on one side from somatic mutation on one side only. Difficulties of these sorts make for overestimation of mutation rates. Further, direct methods assume that all sporadic cases are newly mutant—an assumption apt to be false unless there is strong selection against the trait, or the trait is very rare. Finally, if the sporadic cases are few compared with total cases, illegitimacy may be an important source of error leading to overestimation of mutation rates.

Back mutation is ignored in both methods outlined here. For rare characters, neglect of back mutation probably does not lead to serious error but it may do so in traits (like the Rhesus blood factors, see Vogel 1954b) which are relatively common in most populations in Europe and the United States.

The indirect method assumes population equilibrium between mutation and selection, that is, that the gene frequencies do not change from generation to generation. This assumption is especially dangerous in conditions like the Rh blood factors, where there is selection against heterozygotes (see Haldane 1935, Goodman and Reed 1952).

Lack of full penetrance is not a source of error in the indirect method if we can assume correctly that the heterozygous carrier (in whom the gene is not manifest) has a selective value not different from the normal homozygote. This is not true for the direct method, but the error introduced by incomplete penetrance is not serious if the

undercount of mutants in the offspring generation is balanced by an overcount of an approximately equal number of mutant genes which are not penetrant in the parental generation. A procedure to correct for incomplete penetrance has been suggested by Vogel (1954a).

Special difficulties attach to the problem of estimating relative fitness, f, except when the value of f is considerably less than unity: for example, if none of the affected individuals reproduce, $f = 0$. If fertility is inherited, a bias is introduced when estimates of relative fertility of affecteds are made by pooling sibships of different sizes. As an illustration, achondroplasia is sporadic in about 80 per cent of cases. In such a sporadic condition a sibship of size 10 will contribute 1 comparatively fertile affected and 9 fertile unaffected, while a sibship of size 2 will contribute 1 comparatively infertile affected and *only* 1 comparatively infertile unaffected (Krooth 1953). Special methods for the use of the fertilities of affected individuals and their unaffected sibs in the estimation of fitness have been developed by Krooth (1955) who (1952) also discussed procedures for the measurement of the fertility of parents who are ascertained through their children.

Incomplete location of cases which actually occur in the population, early death of selected genotypes before ascertainment, and improper diagnosis may lead to underestimation of mutation rates. For a fuller discussion of these and related problems see Neel and Schull 1954, Neel 1952, 1954, and Vogel 1954a, especially pp. 328-334.

Data on spontaneous mutation rates for 26 characters are given on page 41. A distinction is made between dominant, recessive and incomplete recessive modes of inheritance because, as shown above, each presents distinctive theoretical problems, given the present state of knowledge, in estimation of mutation rates. In some part, the distinctions may be artificial and reflect incomplete knowledge rather than fundamental differences. It is arbitrary whether "incomplete recessives" are called by that term or by "incomplete dominants." Many human genes once considered to be completely recessive are now known to have some effect in the heterozygous condition (Neel and Schull 1954, pp. 78-83).

Information on a few published estimates has been excluded from the table either (1) because the mode of inheritance is not well established, or (2) available information does not permit proper use of available methods. For example, Haldane's "Porcupine men" (1948)—which, if one will allow high speculation, is the lowest mutation rate (10^{-10}) thus far suggested for man—and Kemp's "dominant Mongolism" (1944)—which, with an equal degree of speculation, is the highest rate—are excluded for the first reason. Mongolism has an incidence in the English population of at least 1.5×10^{-3} (Carter and MacCarthy 1951) which, by the direct method, would give a mutation rate of the order 10^{-3}.

In general, it would seem that the available estimates of spontaneous mutation rates in man are more likely to represent overestimates than the opposite. The rough agreement in the several estimates for different characters where the basic conditions for application of the method used are satisfied approximately gives some ground for supposing that they are of the right order of magnitude. On theoretical grounds we should expect some difference in rates in different populations, especially if their environments are diverse. Muller (1954, p. 11) has speculated that human populations living on the high plateaus of Bolivia and Tibet receive an additional 5 r. per reproductive generation from cosmic radiation, an amount of radiation sufficient to bring about an increase in mutation rate of about 6 per cent.

As Muller (1954) has suggested, it would be possible to make independent checks on some of the available estimates of spontaneous mutation rates in man through

Spontaneous Mutation Rates (Per Gene Per Generation) in Man

No.	Character	Population	Mutant genes per million gametes	Authors
		Autosomal Dominants		
1	Epiloia	England	8-12	Penrose '36
2a	Chondrodystrophy	Denmark	43	Mørch '41
2b	Chondrodystrophy	Sweden	70	Böök '52
2c	Chondrodystrophy	Denmark (mothers over 40 years)	200	Krooth '53
2d	Chondrodystrophy	Denmark	10	Slatis '55
3a	Pegler's nuclear anomaly	Germany, inter al.	100	Pätau & Nachtsheim '36
3b	Pegler's nuclear anomaly	Germany, inter al.	27	Nachtsheim '53
4	Aniridia	Denmark	12	Møllenbach '47
5a	Retinoblastoma	England	14	Philip & Sorsby '44
5b	Retinoblastoma	Michigan	23	Neel & Falls '51
5c	Retinoblastoma	Germany	4	Vogel '54
6	Waardenburg's syndrome	Holland	4	Waardenburg '51
7	Syndactyly	United States	167*	Danforth '23
8a	Palmaris longus muscle	United States (whites)	32*	Danforth '23
8b	Palmaris longus muscle	United States (Negroes)	7*	Danforth '23
9	Lobster claw	England	0.1	Haldane '48
10	Neurofibromatosis	Michigan	100	Crowe, Schull & Neel '55
11	Multiple polyposis colon	Michigan	10-30	T. E. Reed & Neel '55
		Autosomal Recessives		
12	Microphthalmos & anophthalmos	Sweden	10-20	Sjögren & Larsson '49
13	Albinism	Japan	28	Neel et al. '49
14	Total colorblindness	Japan	28	Neel et al. '49
15	Infantile amaurotic idiocy	Japan	11	Neel et al. '49
16	Ichthyosis congenita	Japan	11	Neel et al. '49
17	Cystic fibrosis pancreas	Minnesota	700-1000	Goodman & S. C. Reed '52
18	Epidermolysis bullosa	Sweden	50	Böök '52
19	Amyotonia congenita	Sweden	20	Böök '52
20	Microcephaly	Japan	22-76	Komai '54
21a	Rhesus blood factor	Minnesota	510	Goodman & S. C. Reed '52
21b	Rhesus blood factor	Minnesota	840	Vogel '54b
22	Schizophrenia	Sweden	5000?	Böök '53
		Incomplete Autosomal Recessives		
23	Thalassemia	Sicily	400	Neel & Valentine '47
24	Sickle cell trait	Belgian Congo	1700*	Vandepitte et al. '55
		Sex-linked Recessives		
25a	Hemophilia	England	20	Haldane '35, '48
25b	Hemophilia	Denmark	32	Andreassen '43
26a	Pseudohypertrophic muscular dystrophy	Utah	100	Stephens & Tyler '51
26b	Pseudohypertrophic muscular dystrophy	North Ireland	40-60	Stevenson '53

Maximal estimates are marked with an asterisk (*).

(Modified, with additions, from Neel & Schull 1954, and Nachtsheim 1954.)

studies of linkage of the genes in question with common marker genes such as those controlling the red blood cell antigens.

Known, spontaneous mutation rates per gene per generation in man tend to center about 1 in 50,000 in the more reliable estimates, although rates as high as about 1 in 10,000 are as firmly established as any of the better estimates. Study of a phenomenon as rare as spontaneous mutation in man requires examination of samples much larger than those usually investigated in the medical and human-biologic sciences. Often it is possible to make good use of records collected for other purposes. It is not an accident, nor is it self-interest alone, that we know more about spontaneous mutation rates in man than in any other vertebrate, including the laboratory mouse. Hundreds of thousands of human individuals are given medical examinations each year, and a large volume of results of such examinations are preserved and made available for study. In many cases a relatively small amount of additional information would increase greatly the value of the conventional medical history for genetic investigations. Accurate information on the distribution of hereditary traits in the parents, the siblings and the children of observed individuals is especially needed.

In their routine affairs, clinical orthopaedists may see many cases whose etiology cannot be understood adequately without reference to the genetic factor. Also, orthopaedists are in a favorable position to add much new information to present knowledge of the genetics of congenital and other malformations There is a very wide range of hereditary variation, both normal and abnormal, in the human skeleton. Although known, inherited variations of the muscular system are limited to about half a dozen well-established traits, more hereditary variations of the bone and the joint systems are known than for any other part of the human body with the exception of the eye and the skin (Falls 1953) Of the large number of possible studies of mutation rates for genes concerned with the muscles, the joints and the bones, only 8 have been carried out. The prospects for important work in this area in the future are good.

REFERENCES

Carter, C., and MacCarthy, D.: Incidence of mongolism and its diagnosis in the newborn, Brit. J. Soc. Med. 5:83-90, 1951.

Crowe, F. W., Schull, W. J., and Neel, J. V.: A Clinical, Pathological and Genetic Study of Multiple Neurofibromatosis, Springfield, Ill., Thomas, 1956.

Dahlberg, G.: A new method for determining the mutation frequency in man, Proc. 8th Internat. Congr. Genet., 1948, Hereditas 35(supp.):555-557, 1948 (1949).

Danforth, C. H.: The frequency of mutation and the incidence of hereditary traits in man, Papers 2nd Internat. Congr. Eugen. 1921, 1:120-128, 1923.

Drinkwater, H.: Phalangeal synostosis anarthrosis (Synostosis ankylosis) transmitted through fourteen generations, Proc. Roy. Soc. Med. (Sect. Pathol.) 10:60-68, 1917.

Falls, H. F.: Skeletal system, including joints, *in* Sorsby, A.: Clinical Genetics, pp. 236-286, London, Butterworth, 1953.

Glass, B.: Genetic changes in human populations, especially those due to gene flow and genetic drift, Adv in Genet. 6:95-139, 1954.

Goldschmidt, R. B.: Theoretical Genetics, Berkeley, Univ. California, 1955.

Goodman, H O., and Reed, S C.. Heredity of fibrosis of the pancreas, possible mutation rate of the gene, Am. J Human Genet. 4: 59-71, 1952.

Haldane, J. B. S.. The rate of spontaneous mutation of a human gene, J. Genet. 31: 317-326, 1935.

———. The rate of mutation in human genes, Proc 8th Internat. Congr. Genet., 1948, Hereditas 35(supp.)·267-273, 1948 (1949).

Kemp, T.: Mutation as a cause of disease, Acta path. et microbiol scandinav. suppl. 54:195-208, 1944.

Krooth, R. S . The fertility of the parents of abnormals, Ann. Eugenics 17:79-89, 1952.

———: Comments on the estimation of the mutation rate for achondroplasia, Am. J. Human Genet. 5:373-376, 1953.

———: The use of the fertilities of affected individuals and their unaffected sibs in the estimation of fitness, Am. J. Human Genet 7:325-360, 1955.

Lenz, F.: Über Häufigkeit und Bedeutung von Mutationen in menschlichen Populationen, Homo 3:145-148, 1952.

Li, C. C.: Population genetics, Chicago, Univ. Chicago, 1955.

Mørch, E. T.: Chondrodystrophic dwarfs in Denmark, Opera Ex Domo Biol. Hered. Human. Univ. Hafn. 3:1-200, 1941.

Muller, H. J.: Our load of mutations, Am. J. Human Genet. 2:111-176, 1950.

———: The manner of dependence of the "permissible dose" of radiation on the amount of genetic damage, Acta radiol. 41: 5-20, 1954.

Nachtsheim, H.: Die Mutationsrate menschlicher Gene, Die Naturwissensch. 41: 385-392, 1954.

———: Problems in the estimation of the frequency of uncommon inherited traits, Am. J. Human Genet. 6:51-60, 1954.

Neel, J. V.: The population genetics of two inherited blood dyscrasias in man, Cold Spring Harbor Symp. on Quant. Biol. 15:141-158, 1951.

———: The study of human mutation rates, Amer. Naturalist 76:129-144, 1952.

Neel, J. V., Kodani, M., Brewer, R., and Anderson, R. C.: The incidence of consanguineous matings in Japan, with remarks on the estimation of comparative gene frequencies and the expected rate of appearance of induced recessive mutations, Am. J. Human Genet. 1:156-178, 1949.

Neel, J. V., and Schull, W. J.: Human Genetics, Chicago, Univ. Chicago, 1954.

Popham, R. E.: The calculation of reproductive fitness and the mutation rate of the gene for chondrodystrophy, Am. J. Human Genet. 5:73-75, 1953.

Race, R. R., and Sanger, R.: Blood Groups In Man, ed. 2, p. 383, Oxford, Blackwell, 1954.

Reed, T. E., and Neel, J. V.: A genetic study of multiple polyposis of the colon, Am. J. Human Genet. 7:236-263, 1955.

Slatis, H. M.: Comments on the rate of mutation to chondrodystrophy in man, Am. J. Human Genet. 7:76-79, 1955.

Sorsby, A., ed.: Clinical Genetics, London, Butterworth, 1953.

Vandepitte, J. M., Zuelzer, W. W., Neel, J. V., and Colaert, J.: Evidence concerning the inadequacy of mutation as an explanation of the frequency of the sickle cell gene in the Belgian Congo, Blood 10:341-350, 1955.

Vogel, F.: Über Genetik und Mutationsrate des Retinoblastoms (Glioma retinae), Ztschr. menschl. Vererb.- u. Konstitutionslehre 32: 308-336, 1954a.

———: The mutation rate of the Rh-loci—a critical review, Am. J. Human Genet. 6: 279-283, 1954b.

Wright, S.: Population structure in evolution, Proc. Amer. Philos. Soc. 93:471-478, 1949.

Estimation del Proratas de Mutation in le Homine

Summario in Interlingua

Sequente un breve revista del mutation in general, es delineate duo methodos de estimar le proratas de mutation spontanee in le homine per gen per generation. In le methodo directe le prorata de mutation, u, es obtenite per $u = c(s/t)$, e in le methodo indirecte per $u = c(1 - f)x$, ubi c es un constante pro le modo de hereditate (= ½ pro le dominantes autosomal), s es le numero total de casos sporadic, e t es le numero total de nascentias in un date population e un date periodo de tempore, f es le relative validessa physic de un individuo con un tracto dominante, e x es le frequentia del tracto al nascentia in le population. Exemplos numeric del duo methodos es presentate. In isto le datos de Mørch super achondroplasia in Danmark es usate. Es brevemente discutite alicun premissas necessari pro le correcte application del duo methodos.

Es facite un presentation tabular de 36 studios super le proratas de mutation spontanee de 26 genes. Le estimationes le plus firmemente establit se trova inter 1 per 10.000 e 1 per 50 000 mutationes per gen per generation reproductive.

Con le exception del oculo e del derma, un plus grande numero de variationes hereditari es cognoscite pro le systemas ossee e articular que pro ulle altere parte del corpore human, sed solmente octo studios ha essite executate in re le proratas de mutation de genes que concerne le musculos, le articulos, e le ossos. Le prospectos de importante labores futur in iste campo es bon.

6

Examination of the Possibility That Certain Skeletal Characters Predispose to Defects in the Lumbar Neural Arches

T. D. STEWART, M.D.*

THE PROBLEM

The extraordinarily high incidence of lumbar neural arch defects found in the skeletons of Alaskan natives (Eskimos and Aleuts)—a figure that ranges from approximately 15 to 50 per cent, according to geographic area—offers a unique opportunity for study of the etiology of these abnormalities.[11] In a recent paper based on material from these sources preserved in the United States National Museum, the author[12] showed that the progressive increase in arch defects from early childhood to maturity, together with the accompanying spreading segmental involvement, necessitated abandonment of the view—then held widely—that the defects were hereditary. The substitute explanation advanced by the author was based largely on some unusual postures commonly assumed by the Alaskan natives, which put extra strain on the low back and ultimately on the partes interarticulares of the lower lumbar vertebrae. He felt that long-continued strain at these points could lead to local deossification, ending in fatigue fractures or in accidental fractures from falls on ice.

Such an explanation of the neural arch defects in this particular racial group, which denies heredity and places the cause on environmental factors acting through the mechanics of the body, ignores the possibility that predisposing hereditary factors may exist within the body, and especially in the skeleton. In other words, is it possible that heredity, in the form of skeletal anomalies, still plays an important, though indirect, role among Alaskan natives in causing the defects? The need to answer this question led the author to undertake the present study as a sequel to his earlier publication.

RESEARCH MODEL

The plan of this study has involved the re-examination of much of the same material used earlier in order to set up 2 comparable series of skeletons, 1 with neural arch defects, 1 without. In the new examination the following additional observations were recorded: (1) vertebral formula; (2) nature of transitional lumbosacral vertebra when present (including type of articulation with sacrum, status of fusion and inclination of anterior surface with that of S-1); (3) type of sacrum (hypobasal or homobasal); (4) dimensions of sacrum (including maximum width, transverse and anteroposterior diameters of the superior surface of the 1st centrum, maximum and minimum distances between the superior articular facets of S-1, depth and curvature of the same facets, angle of the superior surface of the 1st sacral centrum with vertical, and angle be-

*Washington, D. C.

tween superior and anterior surfaces of S-1); and (5) dimensions of the last 3 lumbar vertebrae (particularly the vertical distance between the tips of the articular facets and the anterior and the posterior heights of the centra in the mid-line). Explanation of the methods used in making these observations will be deferred until the observations are discussed individually.

As will be seen, these observations are limited to the lumbosacral region. The selection was arrived at after considerable thought and seems to represent the main skeletal characters involved in low back stresses. Although all these observations have been analyzed, limitations of space will permit reporting in detail only those bearing most directly on such important variations as sacral inclination, lumbosacral curvature and lumbosacral articulation. Of course, it must be borne in mind that the observations have been made on dried and disarticulated skeletons. It does not follow that the positioning of the bones during observation was the same as existed in life. However, it may be remarked that, considering the limitations of both gross osteologic and roentgenologic examinations, this study has a bearing on the

TABLE 1. GEOGRAPHIC DISTRIBUTION IN THE MATCHED SERIES

LOCALITY	MALES: With Arch Defects	MALES: Without Arch Defects	FEMALES: With Arch Defects	FEMALES: Without Arch Defects
North of Yukon:				
Point Hope	9	12	5	4
Metlatavik	—	1	—	—
Wales	1	1	5	1
St. Lawrence Island	3	1	3	6
Kowieruk	1	3	1	—
Port Clarence	2	1	—	1
Rocky Point	4	—	5	3
Golofnin Bay	6	3	4	4
Norton Bay	—	1	—	—
Unalakleet	—	—	—	1
St. Michael's Island	—	2	1	—
Pastolik	2	—	—	2
	28	25	24	22
Yukon and South:				
Lower Yukon	2	12	3	2
Hooper Bay	2	4	—	3
Nunivak Island	—	2	—	6
Kuskokwim River	9	—	3	—
Bristol Bay	4	2	3	—
	17	20	9	11
Aleutians and Kodiak:				
Kagamil Island	—	—	2	3
Umnak Island	—	—	1	—
Kodiak Island	—	—	9	9
	—	—	12	12
Total	45	45	45	45

subject of predicting low back troubles, as in pre-employment examinations.

COMPARISONS OF MATCHED SERIES

In order to limit selection in setting up the series of skeletons for comparison, all cases of adults with arch defects were examined in the order in which they had been received and catalogued by the museum. Since many specimens had to be eliminated owing to damage or absence of critical parts, and since females have fewer arch defects than males, only 45 females with arch defects were found to be usable for the present purpose. Accordingly, the male series was limited also to 45—the first 45 examined. Next, the examination was begun in the same way for the cases without arch defects. The male series of 45 thus obtained required no adjustment in order to make it geographically comparable with the other male series; the 2nd female series did require some adjustment. However, the adjustment in the females was made solely on the basis of *geography and without knowledge of the* skeletal characters under consideration.* Table 1 shows that the final series compare reasonably well so far as geographic sources are concerned.

The distribution of ages in the matched series is not so easy to determine, because skeletal age cannot be estimated with cer-

*The adjustment consisted of removing 17 cases derived from the Lower Yukon and replacing them with an equal number from the other 2 main regions: 5 from Rocky Point and St. Lawrence Island, and 12 from Kodiak and Kagamil Islands

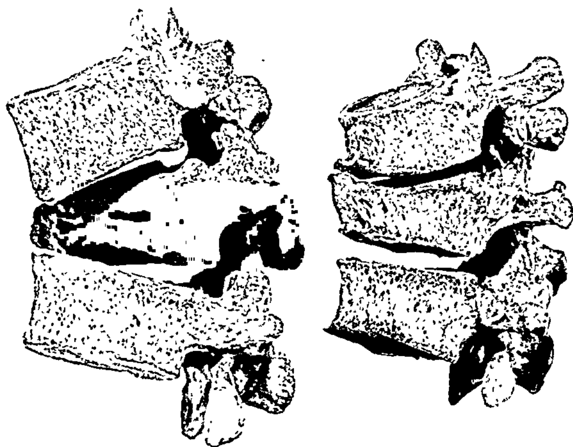


FIG 1. Two examples of collapsed centrum. (*Left*) L-1, male Eskimo from Nunivak Island (U.S.N.M No 339,260). (*Right*) T-11, female Eskimo from St. Lawrence Island (U.S.N.M No 345,051) Both probably are in the age range 30 to 35 years. No arch defects present

tainty after epiphyseal union is completed. For this reason, and following my previous practice,[12] I show in Table 2 the ages expressed in terms of amount of hypertrophic arthritis present. In general, this table shows the series to be fairly well balanced chronologically, with the males showing, if anything, a little more arthritis than the females.

Of somewhat incidental interest is the distribution of arch defects according to sex in the series obtained under these conditions of selection. Table 3, in which a detailed distributional analysis is presented, shows clearly that sex per se is not a factor in determining the type of defect.

Also of incidental interest is the distribution of pathologically deformed vertebral centra throughout the spines used in this study. I refer here to cases in which the anterior height is more or less reduced in one or more centra (Fig. 1). Some of these cases probably represent compression fractures, whereas others possibly were caused by tuberculosis, which has been prevalent among Alaskan natives since contact with whites. Because of my inability in many cases to rate the condition as acute or chronic and to say whether it occurred before, with or after the arch defects, I have simply listed the specimens by sex and series (Table 4). Although some cases may have been overlooked, since this was not one of the standard observations, it appears that, except among the females with arch defects, there is a fairly uniform distribution. Two thirds of the cases listed are from the northernmost part of Alaska. Within the spine the abnormal centra tend to concentrate at the thoracolumbar junction, which will be recognized as the usual site of election for both compression fractures and tuberculosis.

TABLE 2. AGE DISTRIBUTION IN THE MATCHED SERIES

AGE	WITH ARCH DEFECTS			WITHOUT ARCH DEFECTS		
	Males	Females	Total	Males	Females	Total
20-25 years	2	6	8	6	3	9
Over 25 (no arthritis)	7	9	16	10	7	17
Over 25 (slight arthritis)	12	19	31	11	20	31
Over 25 (moderate arthritis)	24	11	35	18	15	33
Total	45	45	90	45	45	90

TABLE 3. SERIES WITH ARCH DEFECTS: TYPE OF DEFECT BY SEX

TYPE	MALES	FEMALES
Single segment involvement:		
Bilateral at pars interarticularis	28	26
Unilateral at pars interarticularis	1	—
Complex*	6	7
	35	33
Multiple segment involvement:		
Bilateral at pars interarticularis	6	11
Complex	4	1
	10	12
Total	45	45

*Some combination of defects involving sites other than or in addition to the pars interarticularis.

TABLE 4. DISTRIBUTION OF PATHOLOGICALLY DEFORMED VERTEBRAE IN MATCHED SERIES

Specimen Number	Location in Spine*												
	T-4	5	6	7	8	9	10	11	12	T-13 L-1	L-2	3	4
With arch defects:													
Males:													
332539†									+	–	–	+	–
333467†				–	+	–				+	–	+	
351248			–	–	–								
351255			+					+					
352394†					+				+				
352403†									+	+			
Females:													
333471†								–					
346013†										–			
346294†									+				
352377†										+			
363547										+	–		
378619‡								–	–	+	–	–	
Without arch defects:													
Males:													
339260										+			
345308	+				–	+			?	+	–	?	
345352									–	+			
346177†												+	–
346192†								+	–	+			
346199†							–	–	–				
346210†								+	+				
Females:													
339115				+			+		+	+			
346004†									–	+			
346010†								+	+	–			
346051†							–	+	–				
366693‡					–					–	+	–	
374635‡				+	–		+		+	+	–	+	–
Total: Pronounced	1	0	1	2	2	1	2	5	8	12	1	4	0
(11–L-1)									25				
Total: Slight	0	0	1	2	4	1	2	3	6	4	6	2	3
(11–L-1)									13				

* A plus sign signifies a segment with pronounced deformity; a minus sign signifies a segment with slight deformity.
† Indicates a locality north of the Yukon River.
‡ Indicates a locality in the Aleutians or on Kodiak Island.

DATA ON U. S. WHITES

During the course of this study the author visited Western Reserve University Medical School in Cleveland and recorded the same set of observations on 29 white male skeletons with lumbar neural arch defects from the Todd Collection. He is indebted to Dr. Mildred Trotter, of Washington University School of Medicine, St. Louis, for a list of

the skeletons in the Todd Collection under 50 years of age with neural arch defects. The skeletons studied were selected from this list; hence they are not a random sample of that part of the white male population with neural arch defects. Nevertheless, this sample will serve as a sort of standard with which to compare the Alaskan natives.

ANALYSIS OF SELECTED OBSERVATIONS

Vertebral Formula

In order to establish the proper relationships of the features under observation in the lumbosacral region, the numbers of segments in each of the component parts of

Table 5. Variation in Numbers of Presacral Segments in Matched Series*

Presacral Segment	With Arch Defects			Without Arch Defects		
	Males	Females	Total	Males	Females	Total
23	—	1	1	1	1	2
24	42	41	83	43	42	85
25	3	3	6	1	2	3
Total	45	45	90	45	45	90

*Unfused lumbosacral transitional vertebrae are treated here as part of the sacrum because of the articulation therewith through one or both lateral processes.



Fig. 2. Variations in the sacrum. (*Left*) Promontory at level of arcuate lines of innominates, hence classified as homobasal. Male, arch defects present (U.S.N.M. No. 346,290, Pt. Hope). (*Right*) False promontory at arcuate level and transitional segment articulating with sacrum on right side only. Female, no arch defects present (U.S.N.M. No. 345,754, Hooper Bay).

the vertebral column were counted. In cases in which segments had been lost or damaged, the exact number present originally could not be determined accurately. When this involved the cervical region, 7 segments were assumed confidently to have existed because of the stability of this region; but when this involved the thoracic region, 12 segments were assumed hesitantly to have been present, solely because the chances greatly favor this modal number rather than the much less common variant numbers 11 and 13. Assumptions regarding the original number of lumbar vertebrae were not entertained, and here all doubtful cases were excluded. In this way a presacral count of segments was arrived at which is summarized for the matched series in Table 5. According to the evidence in this table, a significant difference that can be related to the occurrence of neural arch defects does not seem to exist.

It is interesting to note that 4 of the 29 white males with arch defects examined in Cleveland had 25 presacral segments (13.8%). In the comparable Alaskan series only 3 out of 45 were in this category (6.7%).

Transitional Vertebrae

The footnote to Table 5 calls attention to a fairly common lumbosacral anomaly in which a vertebra—usually the 25th segment—continues to articulate with the sacrum through one or both lateral processes until after completion of ossification of the sacrum proper (Fig. 2, *right*). Each lateral articulation fuses ultimately, and then the condition is often difficult to diagnose, except through the persistence anteriorly of an angle between the upper two centra (Fig. 3, *right*). This angle, which is usually at the level of the iliopectineal, or arcuate, lines on the inner sides of the innominates, repre-

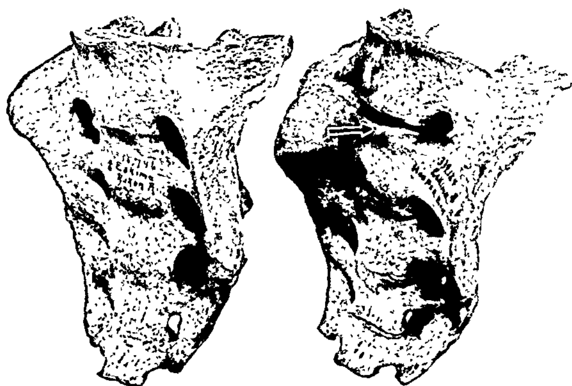


Fig. 3 Variations in the sacrum. (*Left*) Whole first segment above level of arcuate lines of innominates, hence classified as markedly hypobasal. Male, no arch defects present (U S N M. No 345,746, Yukon River). (*Right*) False promontory at arcuate level marked by arrow, transitional segment fused completely on both sides. Male, no arch defects present (U S N M No 339,124, Hooper Bay).

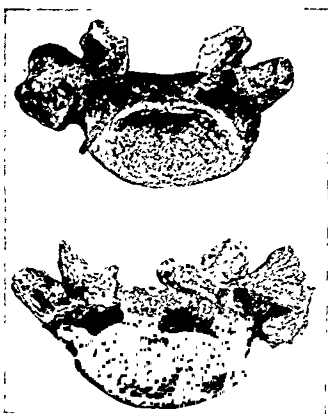


FIG. 4. Transitional vertebrae (unilateral) with arch defects, viewed from below and behind. (*Top*) Articulation with sacrum on left; bilateral arch defects at pars interarticularis (U.S.N.M. No. 363,547, Bristol Bay, female). (*Bottom*) Articulation with sacrum on right; arch defects at pars interarticularis on left and in mid-line (U.S.N.M. No. 346,191, Pt. Hope, male).

sents the promontory of the sacrum at an earlier stage of development and is referred to as the false promontory. In distinguishing between sacra of this type and those (considered in the next section) in which the angle is lacking—hence there is a single promontory at or above the level of the arcuate lines (Fig. 3, *left*)—no causal differences are implied.

The expression *transitional vertebrae* here covers all stages of the anomaly just de-

TABLE 6. CHARACTER OF FIRST SACRAL SEGMENT RELATIVE TO PRESENCE OR ABSENCE OF DEFECTS IN THE LUMBAR NEURAL ARCHES, SEX AND LEVEL IN SPINE

SERIES	VERTEBRAL NUMBER	TRANSITIONAL*	MARKEDLY HYPOBASAL	MED. TO SL. HYPOBASAL	HOMOBASAL
With arch defects:					
	24	—	—	—	—
Males (45)...........	25	11	5	25	1
	26	—	—	2	1
	24	—	—	—	1
Females (45)...........	25	7	8	27	1
	26	—	—	1	—
Total (90)........ ...		18	13	55	4
Without arch defects:					
	24	2	—	1	—
Males (45)	25	7	5	27	—
	26	—	—	2	1
	24	1	—	—	—
Females (45)...	25	6	6	26	4
	26	—	—	2	—
Total (90)...........		16	11	58	5

*A segment that forms an angle with the next [illegible]

scribed. As many writers have pointed out,[1,4,6,14] such vertebrae may be in the process either of liberation from (lumbarization), or incorporation into (sacralization), the sacrum, depending on the direction in which the pelvic girdle is moving on the column. Without going farther here into the broad biologic significance of this anomaly, attention is called to Lanier's statement[4] that "morphologically significant fusion [as distinguished from pathologic fusion] of vertebrae at the lumbosacral level is a complex process related to other intersegmental variations in the spine, and that the process is genetically influenced and determined at an early embryological stage [p. 370]."

Lanier's statement is the conclusion of his argument directed against Thieme's view[13] that fusion between a transitional vertebra and the sacrum is nature's response to the stresses of man's upright posture and thus her effort to "increase the security of the area." I side with Lanier in this argument, and take this opportunity to point out also that the nonupright anthropoid apes have transitional vertebrae that fuse in the same way as in man.[8,9]

Be this as it may, and regardless of whether or not fusion has occurred, transitional vertebrae are less mobile than are typical last lumbar vertebrae. Such being the case, does vertebral stabilization of this kind have any influence on the occurrence of arch defects? So far as our specimens are concerned, Table 6 shows that just about as many transitional vertebrae occur in the series with arch defects as in that without (18 vs. 16). However, it is significant that transitional vertebrae per se, are largely immune from defective arches. In the 18 cases from Alaska in which a transitional vertebra and arch defects are associated in the same spine, the defects occur, with only two exceptions (Fig. 4), in the true lumbar vertebrae instead of in the stabilized vertebrae.

Hypobasality of Sacrum

In the preceding section we have considered a relatively uncommon condition that results in an anomalous type of sacrum—one with a double promontory. By contrast, most sacra have only a single promontory. Yet, even in this modal type of sacrum the level of the promontory varies in relation to surrounding structures (Figs. 2 and 3, *left*).

Radlauer[5] gives a classification of sacra that takes into consideration the relationship of the promontory (his basal plane) to the partes laterales. According to his scheme, when the proximal border of the facies auricularis lies below the basal plane, he speaks of the partes laterales of the sacrum as *hypobasal;* when they are on the same plane, he speaks of them as *homobasal;* and when the proximal border of the facies auricularis lies above the basal plane, he speaks of the partes laterales as *hyperbasal.* Since the "proximal border of the facies auricularis" is too indefinite for the present purpose, I have followed Thieme[13] in using, as a more exact point of reference on each side, the intersection of the arcuate line with the proximal border of the facies auricularis. By this interpretation, only the first 2 categories of Radlauer's classification are needed.

With this explanation, we may ask: Do neural arch defects vary in frequency with the hypobasality or the homobasality of the sacrum? The answer as given by the present study appears in Table 6. As in the case of transitional vertebrae, markedly hypobasal sacra occur about as frequently in the series with arch defects as in that without (13 vs. 11). Likewise, the intermediate grades of hypobasality, which are so common, are represented about equally in the matched series (55 vs. 58). On the other hand, homobasality is so rare as to be of little significance.

Long "Prearcuate" Spines

We now have considered separately three

situations in which the spine above the level of the arcuate lines is increased in length beyond the mode by at least one whole segment: (1) an extra segment in the presacral region; (2) a transitional lumbosacral vertebra; and (3) a sacrum exhibiting marked hypobasality. As considered individually, none of these anomalies seems to have much relation to lumbar neural arch defects, but the little difference that exists tends to favor the series with defects. In view of this trend, we may ask: How much do long "prearcuate" spines predispose in general to lumbar neural arch defects? Combining the figures in Tables 5 and 6, there are 19 males and 18 females, or 37 individuals, in which such spines occur along with arch defects (41%), whereas there are 13 males and 14 females, or 27 individuals, in which such spines do not occur with arch defects (30%). A difference of 11 per cent is impressive and seems to indicate that the long "prearcuate" spine is a contributing factor in the production of arch defects among the Alaskan natives.

By way of contrast, it is interesting to consider the frequency of long "prearcuate" spines in the series of white males with arch defects. Here, as we have seen, an extra presacral segment is of more common occurrence than in the Alaskan natives. On the other hand, the white series contains only one transitional vertebra and only one sacrum with markedly hypobasal lateral parts. This amounts to a frequency of only 20 per cent.

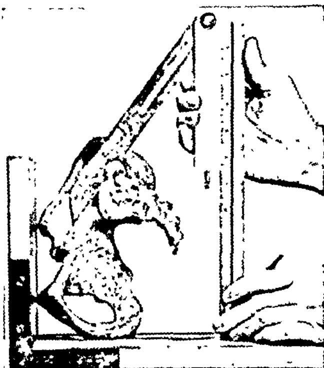


Fig. 5. Method of measuring inclination of top sacral surface in mid-line. Left innominate has been removed for better visibility after orientation of pelvis, which involves contact of both anterior *superior spines and both pubic tubercles* with vertical. Usually the pelvis is held together with rubber bands.

Inclination of Sacrum

According to Schultz,[7] the position of the human sacrum within the pelvis has not changed much as a result of the assumption of erect posture This (and other relationships in the lower parts of the pelvis), he says, "implies that the human trunk became erect only above the sacro-iliac articulation [p 357]." This *interpretation undoubtedly* is correct and explains why the human sacrum presents a variably inclined surface for the support of the presacral vertebral column and its appendages. Largely because of the *inclination* of the sacrum, considerable stress is brought to bear on the parts anchoring the lower lumbar vertebrae to the sacrum and to one another. This is the reason that man alone of all the primates suffers from mechanically produced lumbar neural arch defects. Furthermore, it is the *reason that the inclination of the sacrum* is of interest in the present study.

Given only the dried skeleton, it is not possible to re-establish with certainty the position of the sacrum in life relative to the vertical. The reason is that no two bony points in the pelvis maintain a constant vertical relationship in life *from person to* person, or even probably at different times in the same person (personal communication



FIG. 6. L-5 with bilateral arch defects at pars interarticularis, viewed from behind to demonstrate wedge shape. Note that from this position both superior and inferior surfaces are visible (U.S.N.M. No. 377,709, Kodiak Island, female).

from W. S. Cornwell, based on roentgenograms of the pelvis taken of standing subjects). This being the case, the next best thing is to relate the observations on the sacrum to an arbitrary orientation of the pelvis. Therefore, for the present purpose, I positioned each pelvis so that the anterior superior spines of the ilia and the tubercles of the pubes were in contact with a vertical plane. Figure 5 shows this arrangement, as well as the method used to obtain the angle of inclination of the top surface of the 1st sacral centrum.

As will be seen in Figure 5, the angle of inclination was measured in the mid-line. The surface here usually is far from flat, often being depressed in the middle and raised on either side. Also, occasionally a transverse ridge crosses the median depression, and it is necessary in such cases to see that the arm of the angle is equidistant above the anterior and the posterior borders. Owing to such irregularities, there is some uncertainty that the mid-line represents the true functional slope of the surface. Nevertheless, the angle obtained should be close enough for the present purpose.

Table 7 shows distributions of the angle of inclination by 10° intervals for all cases in the Alaskan series in which S-1 is not of the transitional type. As will be recalled, a transitional vertebra forms an angle anteriorly with the rest of the sacrum. As a result, the top surface of the transitional vertebra is less inclined in relation to the vertical than is the case in normal sacra. The remaining cases have been pooled, regardless of segmental number, because the angle does not seem to depend on length of spine. On this basis the distributions still show a considerable range. Although the females seem to have a little more slope than the males, this may be merely a reflection

TABLE 7. DISTRIBUTION OF THE ANGLE OF INCLINATION OF THE TOP SURFACE OF THE 1ST SACRAL CENTRUM, ACCORDING TO SEX AND PRESENCE OR ABSENCE OF LUMBAR NEURAL ARCH DEFECTS*

Series	Class Interval						Average
	—20°	21-30°	31-40°	41-50°	51-60°	61°+	
Alaskan natives							
With arch defects:							
Males (34)	—	4	12	14	4	—	40.2°
Females (38)	2	8	8	17	2	1	38.4°
Without arch defects:							
Males (36)	—	2	10	19	4	1	43.2°
Females (38)	2	4	13	15	4	—	39.5°
U. S. whites							
With arch defects:							
Males (27)	—	3	10	13	—	1	40.9°

*Cases in which S-1 is transitional in type have been eliminated.

TABLE 8. CONTRIBUTIONS OF LAST 3 LUMBAR VERTEBRAE TO LUMBAR LORDOSIS: AVERAGE HEIGHTS OF CENTRA, ANTERIORLY AND POSTERIORLY, ACCORDING TO SEX AND PRESENCE OR ABSENCE OF LUMBAR NEURAL ARCH DEFECTS*

GROUP	LAST LUMBAR SEGMENT		NEXT TO LAST LUMBAR SEGMENT		SECOND FROM LAST LUMBAR SEGMENT		ALL 3 LUMBAR SEGMENTS		
	Anter. mm.	Post. mm.	Anter. mm.	Post. mm.	Anter. mm.	Post. mm.	Anter. mm.	Post. mm.	Dif. mm.
Alaskan natives:									
With arch defects:									
Males (32)......	24.8	21.1	24.1	23.8	24.0	25.7	72.8	70.5	2.3
Females (34)......	25.3	20.9	24.7	23.8	24.8	25.8	74.9	70.5	4.4
Without arch defects:									
Males (34)......	25.3	22.1	24.3	24.4	24.2	26.3	73.7	72.9	0.8
Females (34)......	24.0	21.4	23.6	23.4	23.9	25.0	71.5	69.7	1.8
U. S. whites:									
With arch defects:									
Males (28)......	28.6	21.8	27.8	26.0	27.6	27.3	84.0	75.2	8.8

*Transitional vertebrae excluded.

of the arbitrary orientation of the pelvis, which probably is affected by sex differences in morphology. But the main point is that, whereas the slopes are slightly less in the series with arch defects than in those without, the differences are not significant statistically.

Table 7 also gives the distribution of the comparable cases in the white series. Judging by this sample, the inclination of the sacrum does not differ appreciably between whites and Eskimos.

LUMBAR LORDOSIS

Like the pelvis, the disarticulated lumbar vertebrae defy attempts to re-establish the orientation that characterized them in life. Indeed, the extensive flexibility of this part of the spine in life makes it difficult even to specify a typical relationship. This being the case, we can study only the characters of the individual vertebrae that are involved in the regional curvature, in this case a lordosis.

As is well known, the centrum of the last lumbar vertebra usually is somewhat wedge shaped, the vertical height being greater anteriorly than posteriorly (Fig. 6). In contrast, the centrum two segments higher up (usually L-3) sometimes is slightly wedge shaped in the opposite direction; that is, the vertical height is greater posteriorly than anteriorly. These differently shaped centra probably are compatible with the lordosis in which they participated during life. Remembering that the lordosis itself is not inherited, we may ask: Do the anterior and the posterior heights of the last three lumbar vertebrae show any relationship to the presence or the absence of neural arch defects at these same levels?

Table 8 shows that when arch defects are present in the Alaskan natives, the combined anterior heights of the last three lumbar centra exceed the combined posterior heights by 2.3 to 4.4 mm. on the average; whereas, when defects are not present, these heights differ only by 0.8 to 1.8 mm. The differences would be larger if the wedging present in the last 2 segments, and par-

TABLE 9. METRIC CHARACTERISTICS OF THE SUPERIOR ARTICULAR FACETS OF S-1 ACCORDING TO SEX AND PRESENCE OR ABSENCE OF LUMBAR NEURAL ARCH DEFECTS

GROUP	MAXIMUM DISTANCE BETWEEN FACETS (1) mm.	MINIMUM DISTANCE BETWEEN FACETS (2) mm.	RATIO: $\frac{2 \times 100}{1}$	MEAN DEPTH OF FACETS (3) mm.	RATIO: $\frac{3 \times 100}{\text{MEAN OF 1 \& 2}}$
Alaskan natives:					
With arch defects:					
Males (43)......	51.3	28 4	55.5	0.9	2.2
Females (44)......	45.6	25.1	55.0	0 8	2.3
Without arch defects:					
Males (44)......	51.2	26.9	52.4	1.0	2.6
Females (41)......	47.8	25.8	54.0	0.9	2.6
U. S. whites:					
With arch defects:					
Males (29)......	54.8	30.7	56.0	0 9	2.1

ticularly in the last, was not offset by the counterwedging present in the segment second from the last. Thus, by contrast with the U. S. white males with arch defects, in which the wedging is in the same direction in all three segments, the difference in combined anterior and posterior heights amounts to 8.8 mm. on the average. Although the degree of lordosis present in life depends on other things besides the shapes of the centra,

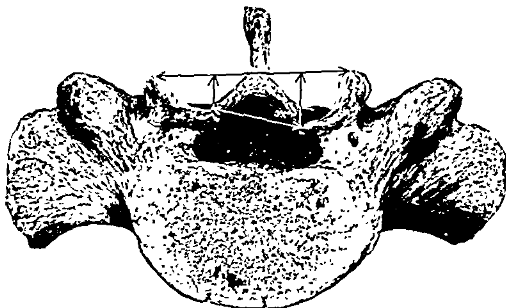


FIG. 7 View of sacrum from above to show measurements relating to articular facets Case selected to show extreme asymmetry of facets. Curvature of left facet near maximum observed. Male, no arch defects present (U.S N.M. No. 333,468, Wales)

TABLE 10. CURVATURE OF SUPERIOR ARTICULAR FACETS OF S-1 ACCORDING TO SEX AND PRESENCE OR ABSENCE OF LUMBAR NEURAL ARCH DEFECTS

Group	Curvature of Both Facets			Side of Greatest Curvature in Asymmetries†	
	None to Slight %	Medium to Pronounced %	Marked Asymmetry* %	Right %	Left %
Alaskan natives:					
With arch defects:					
Males (43)	83.7	11.6	4.6	9.3	7.0
Females (45)	86.7	8.9	4.4	4.4	26.7
Without arch defects:					
Males (44)	63.6	20.4	15.9	18.2	31.8
Females (44)	70.4	25.0	4.5	8.9	20.0
U. S. whites:					
With arch defects:					
Males (28)	85.7	7.1	7.1	10.7	21.4

*Cases in which 1 facet is in the none-to-slight category, whereas the other is in the medium-to-pronounced category.
†All asymmetries regardless of degree.

the figures in Table 8 suggest that the Alaskan natives have flatter backs than whites, and that those without arch defects are the flattest of all. This may mean that increasing lordosis helps predispose to arch defects.

I have not tested the metrical differences shown in Table 8 for statistical significance, because, if such significance can be demonstrated, it may be indicative of things other than sampling characteristics. For instance, in cases of pronounced hypertrophic arthritis, alterations of the superior or the inferior surfaces of the centra may occur, with resultant changes in height. I cannot be sure that all abnormal cases of this sort have been eliminated. Furthermore, transitional vertebrae with small lateral areas of articulation with the sacrum are like typical last lumbar vertebrae, so far as wedging is concerned, and yet have been treated here as 1st sacrals. Thus, the elimination of so many transitional vertebrae from measurement and the substitution of the atypical vertebrae next above probably make the Eskimo spines seem flatter than they are.

LUMBOSACRAL FACETS

The mobilty of the last lumbar segment depends in part on the positions of the lumbosacral facets. As the positions of these surfaces approach the coronal plane, more motion is possible. At such an extreme the facets of the lumbosacral joint offer a contrast with those of the upper lumbar vertebrae, which usually approach the sagittal plane in position and form joints of a type that has been likened by Davis[2] to a carpenter's mortice. Since lumbar neural arch defects are concentrated at the lower end of the lumbar column, where the mortice joint is not much in evidence, it is important to compare the type of joint here with the presence or the absence of arch defects.

In this study, observations on the facets were limited to those of the sacrum that enter into the lumbosacral joint. Figure 7 defines the three measurements taken. In addition, when the articular surface of a facet was not flat, the curvature was judged visually to be slight, medium or pronounced. The specimen shown in Figure 7 was selected

to show both asymmetry and the greatest degree of curvature (pronounced) encountered.

Table 9 gives the averages of the measurements and of two ratios derived therefrom. So far as the matched series of Alaskan natives is concerned, the main difference (not significant statistically) appears in the ratios formed by the depth of the facets in relation to the mean distance between them. For the series with arch defects this ratio is 2.2 to 2.3, whereas for the series without defects this ratio is 2.6. Very little contrast is afforded by the U. S. white males with arch defects: they differ mainly in having greater distances between the facets.

Finally, Table 10 shows that the superior articular facets of the sacrum tend to be more curved and asymmetrical in the series without arch defects than in those with the arch. Thus, the last two tables suggest that a reduction in curvature and relative depth of the articular facets may help increase the frequency of neural arch defects. The tendency for the right facet to be less curved than the left seems to be part of the pattern of sidedness.

DISCUSSION

In the foregoing comparison of matched skeletal series of Alaskan natives, perhaps the outstanding impression centers in the small size of the differences Over and over again in presenting particular observations the conclusion was reached that a difference in this respect between the series with and without arch defects either did not exist or was small and perhaps not statistically significant. Nevertheless, the individual small differences involve rather consistently a pattern of features favoring the increase of mechanical strain in the lower back. Thus, we find neural arch defects associated a little more commonly with (1) a long "prearcuate" spine, which represents a progressive trend in the elongation of the human trunk; (2) an acutely inclined top sacral surface, which, by being contrary to the evolutionary trend, adds to the instability of the lumbosacral area; (3) increased lumbar lordosis, which is a secondary adjustment to erect posture; and (4) reduced depth and curvature of the superior sacral articular facets, which increase the mobility (instability ?) of the lumbosacral junction. However, in reality such a pattern, if it exists, is little more than a statistical concept. The individual elements of this pattern are somewhat unusual in occurrence and are not necessarily associated in a single individual, even when that individual has developed neural arch defects. Indeed, it should be quite evident that the series without arch defects also contains many individuals with one or more of the strain-related features. Stated in another way, the predictive value of these anomalies is low, if not nil, in so far as their use is concerned in foretelling the development of neural arch defects.

Obviously it was impossible to include in the comparison the structure of the bone at the sites of the defects—the partes interarticulares of the neural arches. Therefore, we do not know whether or not weakness of these parts was a factor leading to the occurrence of the defects. Yet, judging from the other skeletal characters here examined, it seems unlikely that the bone at these sites was much, if any, different in the cases that were to develop defects than in those that never did.

Probably structural variability of this sort does not assume more importance as a predisposing factor in causing arch defects because the advantage or the disadvantage that it provides is small. In most circumstances even a seemingly disadvantageous structure will not give rise to this type of disability. On the other hand, when mechanical stresses become sufficiently intense or persistent, bone dissolution ensues, the conformation of the parts making little difference, except to help focus the strain at the point at which the defect occurs This argument is supported by the comparison of Alaskan natives and U. S. whites. Neither group is very different so far as the features under

examination are concerned, yet the vastly higher incidence of arch defects (and of pathologically deformed vertebral centra) among the Alaskan natives indicates that they have been subjected to a precipitating stress factor of greater magnitude.

Up to this point we have been considering heredity in the narrow sense of the skeletal variations exhibited within one species, in this case Homo sapiens. But in a broader biologic sense such skeletal anomalies are expressions only of a pattern of inheritance that make man unique among all primate species. We alone as a species have true upright posture, and, in varying frequencies, arch defects. There can be little doubt that the two are related. We have inherited the basic predisposition to arch defects along with the mechanism for upright posture. In other words, arch defects are a consequence of man's still imperfect skeletal adjustment to upright posture. Only in such a broad sense does heredity play an important role in connection with this disability.

Finally, it should be noted that the present findings were anticipated by certain roentgenographic studies, such as that by Splithoff [10] However, agreement was not foreseeable because of the different approaches used, the different racial groups examined, and the fact that the present study concentrates on a well-defined clinical entity, whereas the others make no distinction between the various disorders classified as "backaches."

REFERENCES

1. Adolphi, H.: Ueber die Variationen des Brustkorbes und der Wirbelsäule des Menschen, Morph. Jahrb. 33:39-86, 1905.
2. Davis, P. R.: The thoraco-lumbar mortice joint, J. Anat. 89:370-377, 1955.
3. Kühne, K.: Die Vererbung der Variationen der menschlichen Wirbelsäule, Ztschr. Morph. u. Anthropol. 30:1-221, 1932.
4. Lanier, Raymond R.: Some factors to be considered in the study of lumbosacral fusion, Am. J. Phys. Anthropol. 12:363-371, 1954.
5. Radlauer, Curt: Beiträge zur Anthropologie des Kreuzbeines, Morph. Jahrb. 38:323-447, 1908.
6. Rosenberg, E.: Ueber die Entwicklung der Wirbelsäule und das Centrale carpi des Menschen, Morph. Jahrb. 1:83-197, 1876.
7. Schultz, Adolph H.: The skeleton of the trunk and limbs of higher primates, Human Biol. 2:303-438, 1930.
8. ———: Growth and development of the orang-utan, Carnegie Inst. of Washington Pub. No. 525 (Contrib. Embryol., No. 182), pp. 57-110, 1941.
9. ———: Age changes and variability in gibbons: a morphological study on a population sample of a man-like ape, Am. J. Phys. Anthropol. 2:1-129, 1944.
10. Splithoff, Clarence A.: Lumbosacral junction: roentgenographic comparison of patients with and without backaches, J.A.M.A. 152:1610-1613, 1953.
11. Stewart, T. D.: Incidence of separate neural arch in the lumbar vertebrae of Eskimos, Am. J. Phys. Anthropol. 16:51-62, 1931.
12. ———: The age incidence of neural-arch defects in Alaskan natives, considered from the standpoint of etiology, J. Bone & Joint Surg. 35A:937-950, 1953.
13. Thieme, Frederick P.: An anatomical relationship predisposing to lumbosacral fusion, Am J. Phys. Anthropol. 9:149-158, 1951.
14. Willis, Theodore, A.: The lumbosacral vertebral column in man, its stability of form and function, Am. J. Anat. 32:95-123, 1923.

Critica del Possibilitate que Certe Characteristicas Skeletal Predispone a Defectos in le Lumbar Arcos Neural

Summario in Interlingua

Reliquias skeletal de nativos de Alaska —eschimos e aleutas—monstra le plus alte cognoscite frequentia de defectos del lumbar arcos neural. Secundo le areas geographic in question, iste frequentia amonta a 15 e usque a 50 pro cento Proque il es hodie

un ben establite facto que le majoritate del defectos de arco non es congenite sed se disveloppa in le curso del vita del individuo, le material de Alaska provide un unic opportunitate pro le studio del etiologia de tal defectos. In iste connexion le autor del presente articulo ha suggerite in un previe publication que le inusual posturas que es communmente prendite per iste nativos es *possibilemente capace a explicar in grande* mesura lor alte frequentia de defectos de arco. Per iste posturas un tension additional es imponite super le dorso inferior e ultimemente super le partes interarticulari del vertebras infero-lumbar. Remane a clarificar le question si hereditate es involvite indirectemente, in le forma de anomalias skeletal que constitue un predisposition a tal defectos.

Le collectiones del Statounitese Museo National ha providite duo geographicamente comparabile series pro le presente studio. Un del series ha defectos, le altere non. Es includite 45 masculos e 45 femininas in cata un del series. Pro objectivos de comparation le Collection Todd del Universitate Western Reserve provideva 29 masculos blanc con defectos de arco. Observationes in iste series esseva limitate a certe variationes concernite con inclination sacral, curvatura lumbosacral, e *articulation lumbosacral. Tal anomalias* occurre solmente un pauco (si del toto) plus frequentemente in le serie con defectos de arco que in le serie sin tal defectos. Tamen, le minime differentias notate es omnes de natura a tender verso un augmento del tension mechanic in le dorso inferior. Es a concluder que nulle del factores studiate ha un valor predictive. In iste senso le studio es de interesse ab le puncto de vista de examines pre-occupational.

7

Anomalies of the Lumbar Spinal Cord and Nerve Roots

R. T. McElvenny, M.D.*

The frequency with which the lumbar area of the spinal cord is explored for extradural lesions makes embryologic variations in this region a matter of interest.

DEVELOPMENTAL IMPLICATIONS

The entire nervous system, except the olfactory epithelium and parts of certain ganglia, is derived from an elongated plate of thickened ectoderm, the *neural plate.* This future nervous system extends longitudinally without sharp demarcation into the thinner nonneural ectoderm. The lateral edges of the *neural plate* curve dorsally to form the *neural folds.* These folds meet and fuse to form the *neural tube* and its central canal. Later the *neural tube* separates from the overlying nonneural ectoderm, which then completely overlies this *neural tube.*

The *neural tube* gradually differentiates into four zones, the floor zone, the roof zone and two lateral zones. The lateral zones, or plates, thicken greatly, and these signify the

*Chicago, Ill.

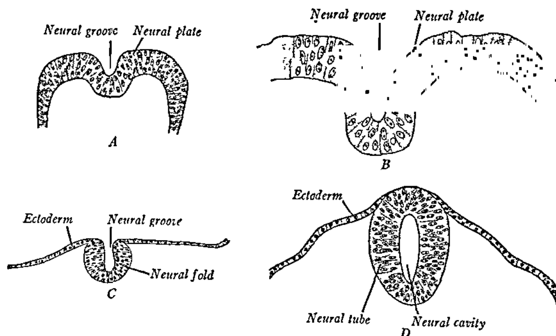


Fig. 1. Four sections showing the development of the neural tube in human embryos. (A) An early embryo (Keibel). (B) At 2 mm. (Grob-Spec.). (C) At 2 mm. (Mall). (D) At 2.7 mm. (Kollmann). (Arey: Developmental Anatomy, Philadelphia, Saunders)

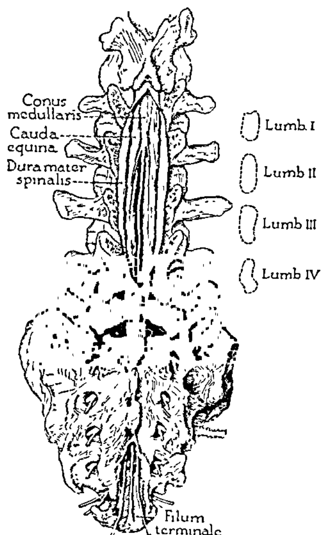


FIG. 2 Normal appearance of lower spinal cord. (Anson, Barry J : Atlas of Human Anatomy, Philadelphia, Saunders)

bilateral character of the *neural tube*. Later, these lateral zones are squeezed transversely by the sulcus limitans into a dorsal and a ventral portion. This process eventually produces a general grouping, so that the dorsal portion of the adult cord becomes mainly receptive in character, and the ventral portion becomes mainly motor in nature (Fig. 1).

During the early development the spinal cord extends from the medulla to the lower end of the sacrum. The bony elements of the spine grow more rapidly than the cord, so that in adult life the spinal cord terminates at about the level of the 1st lumbar vertebra. Below the termination of the spinal cord the lower spinal nerves have had to elongate to accommodate to the growth of the bony column. These nerves descend as the *cauda equina* in the dural sheath to reach their respective exits. The spinal cord itself, having been pulled cephalad by bony growth, leaves a gradually diminishing termination, *the conus medullaris*. This conus eventually narrows into the *filum terminale*, which ends at about the level of the 2nd sacral vertebra. Here the filum perforates the dural sac to continue as a dural sheathed *filum durae matris* to fix firmly into the posterior surface of the coccyx (Fig. 2).

In transverse section the cord shows a centrally placed gray substance, which forms a continuous deeply notched H-shaped column extending the entire length of the cord. The deep ventral median sulcus and the

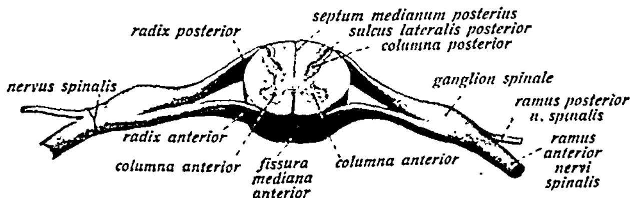


FIG. 3 Diagrammatic representation of a section of spinal cord with nerve roots. (Sobotta-McMurrich Atlas of Human Anatomy, New York, G E. Stechert & Co)

dorsal median sulcus divide the cord into almost complete halves. These halves are joined together by a thin median bridge or commissure. In each half of the cord the gray substance shapes itself into the dorsal and the ventral horn, cornua or column. The gray substance is completely surrounded by the white substance, which is composed of myelinated fibers (Fig. 3).

The spinal nerve roots emerge from the dorsal and the ventral columns and are so named. These fuse into a mixed spinal nerve contained in a dural slip, or sleeve. The spinal nerves leave the bony canal by way of their foramina to connect the various parts of the body to the central nervous system. There are 31 pairs of these spinal nerves. Five pairs are lumbar and five pairs are sacral.

Injury to a dorsal root produces complete loss of sensation and loss of reflexes initiated by sensory stimulation. Overlapping is prevalent, so that unless more than one dorsal root is affected, anesthesia may not be marked.

The cutting of a ventral root produces a complete flaccid paralysis of the muscle or the portion of muscle supplied by the nerve and the loss or the diminishing of the deep reflex controlled by that muscle.

Destruction of a mixed spinal nerve produces both the dorsal and the ventral root effect. When preganglionic visceral fibers are associated with nerve roots, as in the dor-



FIG. 4. (*Left*) Filling of basal and dural spaces in infant with spina bifida and hydrocephalus. (*Right*) Area of lesion shown at left. Note that cord and cauda equina cease in region of arrow. Note below this area that no posterior elements of spinal column are present, even in sacral region.

lumbar region is usual in marked spina bifida. The degree of this affliction varies, so that the gamut may run from no involvement of the cord to premature termination or complete absence of the lumbar cord (Fig. 4).

True Partial Doubling of the Cord (Diplomyelia)

This condition is most common in the lumbar region. Often there is a separation for a segment or two, and then the separated portions fuse together again in the lower lumbar region or before termination. This condition is characterized by two central canals and two posterior "median" septa. In some of these cases this separation is accompanied by the intrusion of a fold of membrane between the two parts. This membrane may be fibrous in nature and contain connective tissue, or even pieces of cartilage and bone. In others the separating membrane is a fold of dura mater. Usually these folds arise ventrally and proceed in a dorsal direction to attach, at times, to the posterior elements of the vertebrae.

The cause of this condition is not clear. In some instances no cause is evident. The abnormality may result from some defect in the cells of the medullary tube. Other cases seem to indicate a local doubling of the medullary tube. This reasoning is based upon the regularity of the outer cornua's being more perfect than the inner, more dorsal, cornua and the continuity of the two central canals.

Theodor's[15] case is of special interest because this was a complete separation of each cord with two separate canals and four anterior and four posterior cornua. The nerve roots came mainly from the outer halves, but the inner, more dorsally situated, cornua had also anterior and posterior root connections (Fig. 5).

In cases of doubling of the cord, the space occupied by the cord is much larger than is demanded by the normal structure. In opening the canal, the membranes bulge

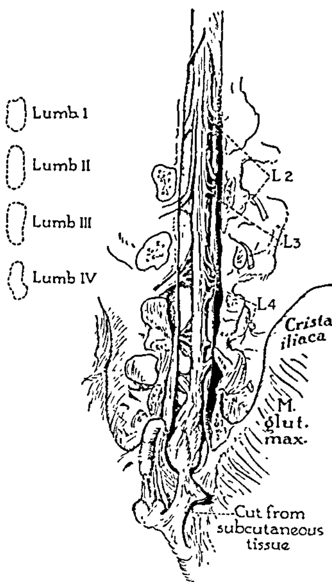


Fig. 6. Sacral cord in an adult. (Anson, Barry J.: Atlas of Human Anatomy, Philadelphia, Saunders)

more markedly, and the canal is larger than normal, with the membranes completely filling the canal.

Partial Doubling of the Lumbar Cord Due to Cartilaginous or Bony Wedges (Diastematomyelia)

Pressure phenomena due to bony or cartilaginous wedges arising from the anterior (ventral) portion of the canal were reported by Cruveilhier[4] over a century ago. Bonome,[2] Sulzer[14] and Bruce[3] also report similar findings. This separation of the cord might conceivably be due to pressure phenomena during the developmental stages.

sal and the upper lumbar regions, vasomotor and trophic changes also are manifest in the skin supplied by the injured nerves.

ABNORMALITIES OF THE LUMBAR CORD

The literature records many marked anomalies of the lumbar cord. Usually these defects are associated with monsters or other marked deviates. Unfortunate as these variations are, they are not subtle. However, the lumbar cord is the seat of many variations that are not so evident. The author was able to collect from the literature between 1880 and 1910 22 cases of proved lumbar cord abnormality in the adult. These people ranged from 17 to 76 years of age. In none had the spinal cord abnormality contributed to the death of the individual. In some of these cases deformity of the feet was noted. This deformity was referred to commonly as cavus. In others a diagnosis of "awkward gait," "poor co-ordination of the legs" or "Friedreich's ataxia" had been made previous to death. Knee jerks, when mentioned, were noted as absent or depressed, or, at other times, as active or brisk. It was impossible to correlate the pathology with the meager physical data.

Spina Bifida

Nerve splitting of the spinal cord in the

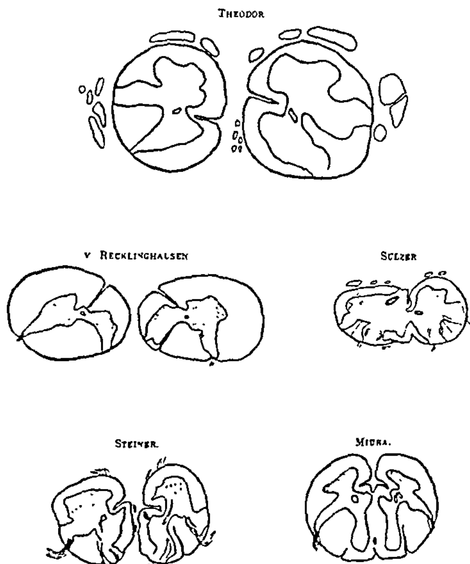


Fig. 5 Various findings in lumbar cord anomalies.

lumbar region is usual in marked spina bifida. The degree of this affliction varies, so that the gamut may run from no involvement of the cord to premature termination or complete absence of the lumbar cord (Fig. 4).

TRUE PARTIAL DOUBLING OF THE CORD (DIPLOMYELIA)

This condition is most common in the lumbar region. Often there is a separation for a segment or two, and then the separated portions fuse together again in the lower lumbar region or before termination. This condition is characterized by two central canals and two posterior "median" septa. In some of these cases this separation is accompanied by the intrusion of a fold of membrane between the two parts. This membrane may be fibrous in nature and contain connective tissue, or even pieces of cartilage and bone. In others the separating membrane is a fold of dura mater. Usually these folds arise ventrally and proceed in a dorsal direction to attach, at times, to the posterior elements of the vertebrae.

The cause of this condition is not clear. In some instances no cause is evident. The abnormality may result from some defect in the cells of the medullary tube. Other cases seem to indicate a local doubling of the medullary tube. This reasoning is based upon the regularity of the outer cornua's being more perfect than the inner, more dorsal, cornua and the continuity of the two central canals.

Theodor's[15] case is of special interest because this was a complete separation of each cord with two separate canals and four anterior and four posterior cornua. The nerve roots came mainly from the outer halves, but the inner, more dorsally situated, cornua had also anterior and posterior root connections (Fig. 5).

In cases of doubling of the cord, the space occupied by the cord is much larger than is demanded by the normal structure. In opening the canal, the membranes bulge

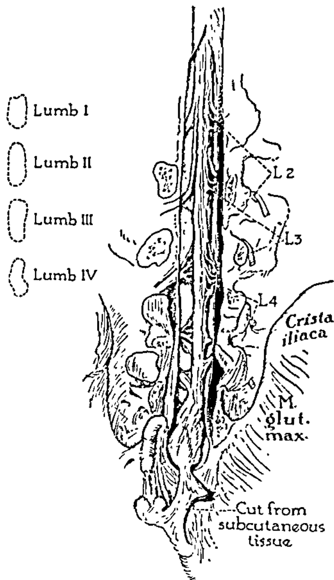


FIG. 6 Sacral cord in an adult. (Anson, Barry J.: Atlas of Human Anatomy, Philadelphia, Saunders)

more markedly, and the canal is larger than normal, with the membranes completely filling the canal.

PARTIAL DOUBLING OF THE LUMBAR CORD DUE TO CARTILAGINOUS OR BONY WEDGES (DIASTEMATOMYELIA)

Pressure phenomena due to bony or cartilaginous wedges arising from the anterior (ventral) portion of the canal were reported by Cruveilhier[4] over a century ago. Bonome,[2] Sulzer[14] and Bruce[3] also report similar findings. This separation of the cord might conceivably be due to pressure phenomena during the developmental stages.

This anomaly may be similar to diplomyelia, but, instead of soft tissue partitions, cartilage and bone are substituted.

Complete Sacral Cord

This portion of the cord is present in the fetus, but the entire cord is drawn cephalad as the bony column outstrips the cord in growth. Anson[1] reports a complete sacral cord in an adult. No history previous to death in relation to physical findings is known of this subject (Fig. 6).

MALFORMATION OF LUMBAR SPINAL ROOTS AND SLEEVES

In describing the variations in the lumbar cord, no mention except by Theodor included the pattern of the nerve roots or the spinal nerves in the affected areas. The developmental abnormalities in the lumbar cord can and do affect the distribution, the size, the location and the pattern of various nerve roots leaving the spinal canal. The cord also may be normal, and variations in the pattern of the nerve root or the spinal nerve may be the only malformation present.

Pathologists, such as Zagoni,[16] have described the various patterns of the lumbar nerve roots, but Fineschi[7] and Ethelberg,[6] publishing in the same year, have tied up clinical data with the operative findings. Ethelberg and Riishede show five variations of the lumbar spinal roots, and their illustrations are shown here. These cases were found in a series of 1,162 laminectomies for pain simulating lumbar disk syndrome. In none of these five cases was a disk protrusion found (Figs. 7-9).

Ethelberg remarks that these abnormally large roots occupy the entire and limited intraspinal space, so that any diminution of this space by even a slight narrowing of the intervertebral disk may result in compres-

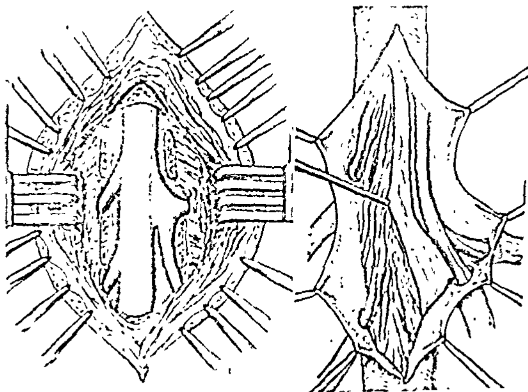


Fig. 7 (*Left*) A dichomatous filum. (*Right*) An abnormal dural sac containing the 5th lumbar and the 1st sacral roots with arachnoidal adhesions between the roots of the cauda equina (Ethelberg & Riishede: J. Bone & Joint Surg. 31B:442-446)

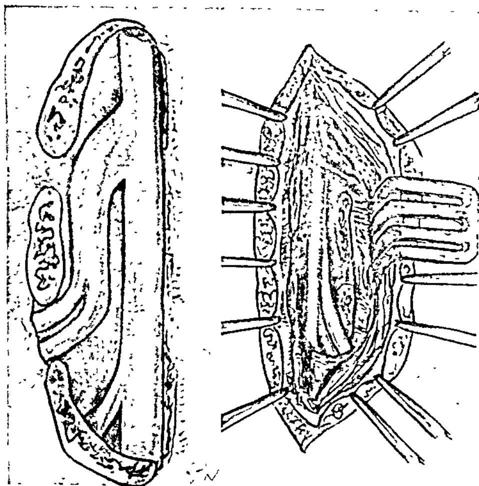
sion of the nerve roots. In these cases neurologic findings were more marked than in the disk syndrome, except that straight leg raising was commonly negative. Otherwise, the histories were indistinguishable from those of intervertebral disk protrusion.

Deyerle and May[5] report a case of disk protrusion with an anomalous nerve root. This case really exhibits an accessory nerve



FIG. 8 (*Opposite*). A plexiform connection between the 5th lumbar and the 1st sacral nerve roots and caudally a thin 2nd sacral nerve root. (Ethelberg & Riishede: J. Bone & Joint Surg. 34B:442-446)

FIG. 9 (*Below*). (*Left*) A common dural sheath for the 5th lumbar and the 1st sacral roots which split up caudally into separate roots, both passing through the 5th intervertebral foramen. (*Right*) An abnormal offshoot of the 5th lumbar nerve root which is considerably enlarged. This root passes through the 5th lumbar intervertebral foramen, together with the 1st sacral nerve root. (Ethelberg & Riishede: J. Bone & Joint Surg. 34B:442-446)



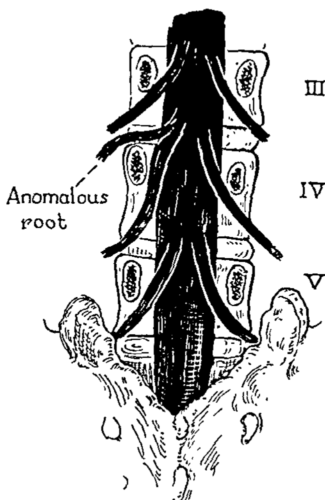


FIG. 10. Illustrates an accessory nerve root. (Deyerle & May: *In* Clinical Orthopaedics, No. 4, Philadelphia, Lippincott)

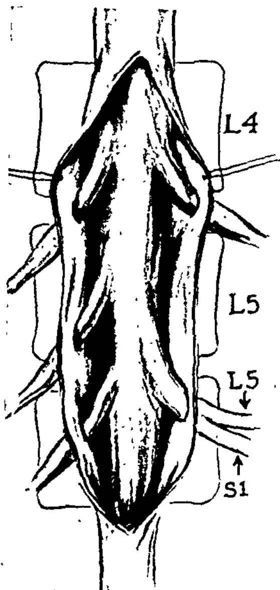


FIG. 11. Illustrates author's case of a dichomatous filum.

root in that the 5th nerve root has an accessory branch (Fig. 10). These authors do not state whether or not the dura mater was opened, so that the true relations of the root above and below were observed.

The author presents one case of a dichomatous filum on the right at the interspace between the 5th lumbar vertebra and the sacrum. This patient had before operation a severe right-sided sciatica. There was diminished sensation over the lateral side of the right foot, a decreased Achilles reflex and marked weakness of the long extensor of the great toe, with diminished sensation over the dorsum of the right great toe. So marked were these findings that two disk lesions were anticipated, one at the 4th interspace and one at the 5th interspace. A myelogram was inconclusive because of the fragmentation of the opaque column. At operation a large protruding disk was found at the bifurcation of the abnormally large root, with most of the disk substance compressing the 5th root. It was exceedingly difficult to mobilize this root, so that the disk substance was removed, beginning on the left. Following mobilization and removal of the disk substance, the dura was opened, and the area shown in the drawing was explored completely. The findings are shown (Fig. 11).

DISCUSSION

The existence of bony encroachment upon the cord or the nerve roots or the presence of a tight (short) filum terminale, a cyst, a transverse constriction band or a tumor pressing upon the cord is common knowledge.

It would be impossible to describe all abnormalities of the spinal cord and the nerve roots, because, as elsewhere, it is almost impossible to list congenital defects. It would appear that classification of congenital abnormalities of the spinal cord and the nerve roots should be made on the basis of obvious and subtle. An attempt has been made to call attention to some of the subtle variations in the lumbar spinal cord and nerve roots.

The embryologic development of the spinal cord signifies its bilateral character. Thus, the variations in the cord itself usually are bilateral. It is singular that in each case of anomaly found in nerve roots the variation has been unilateral.

The resection of a nerve root is not a major disaster, but it is one to be avoided unless done with full knowledge of what one is cutting. The implication in cutting an accessory nerve root (Fig. 10) is different from that of cutting a dichomatous filum (Fig. 11).

An unexplained foot deformity with resistance to correction or an abnormal gait with no true pattern of known etiology should make one consider a lumbar cord abnormality.

SUMMARY

A review of a portion of the literature concerning lumbar cord variations is presented. In this review the most fruitful work was published on this subject bewteen the years 1842 and 1910.

The literature concerning the clinical manifestations of anomalous nerve roots has been reviewed. These anomalies may give rise to symptoms of disk syndrome with or without a herniation of the nucleus pulposus.

One case of spina bifida and one case of dichomatous spinal nerve root have been presented by the author.

REFERENCES

1. Anson, Barry J.: Atlas of Human Anatomy, p. 230, Philadelphia, Saunders, 1950.
2. Bonome, A.: About a rare case of partial doubling of the spinal cord, Arch. Scienze Med., Referat. Neurol. Centralbl., vol. 7, 1888.
3. Bruce, Alexander, and Pirie, Harvey: A second case of partial doubling of the spinal cord, Rev. Neurol. & Psychiat. 4: 6-19, 1906.
4. Cruveilhier, J.: Anatomie pathologique, vol. 1, 1829 and vol. 2, 1842, Paris, Bailliere.
5. Deyerle, W., and May, V., Jr.: Sciatica—etiology and treatment *in* Clinical Orthopaedics, No. 4, pp. 166-179, Philadelphia, Lippincott, 1954.
6. Ethelberg, Sven, and Rüshede, John: Malformations of lumbar spinal roots and sheaths in the causation of low back ache and sciatica, J. Bone & Joint Surg. 34B: 442-446, 1952.
7. Fineschi, C.: Anatomical anomalies of the lumbar spinal roots, Arch. Putti 2: 222-236, 1952.
8. Miura: The genesis of the hollow part in the spinal cord, Virchows Arch. Path. Anat., vol. 117, 1889.
9. Arey, B.: Developmental Anatomy, p. 300, Philadelphia, Saunders, 1917.
10. Rechlinghausen, V.: Studies about spina bifida, Virchows Arch. Path. Anat., vol. 105, 1886.
11. Steiner: About Doubling of the Spinal Cord, Inaugural Dissertation, Koenigsberg, 1895.
12. Stewart, Purves, and Bernstein, Julius: Partial doubling of the spinal cord, Rev. Neurol. & Psychiat. 4:729-736, 1906.
13. Strong, Oliver, and Elwyn, Adolph: Human Neuro-Anatomy, Baltimore, Williams & Wilkins, 1943.
14. Sulzer, Paul: A case of spina bifida connected with bifurcation and doubling of the spinal cord, Ziegler's Beiträge, vol. 12, 1893.
15. Theodor: A case of spina bifida with doubling of the spinal cord, Arch. Kinderh, vol. 24, 1898.
16. Zagoni, C.: Report on an unknown type of nerve anastomoses of the spinal roots, Atti della Società Medico-chirurgica di Padova, 27, 1949.

Anomalias del Medulla e del Radices Nervose Lumbar

Summario in Interlingua

Es presentate un revista de un portion del litteratura concernite con variationes del medulla lumbar. Le plus fertile studios esseva publicate inter 1842 e 1910.

Es presentate un revista del litteratura concernite con le manifestationes clinic de anomale radices nervose. Le litteratura pare indicar que iste anomalias pote producer symptomas de dyndrome de disco, con o sin herniation del nucleo pulpose.

Es generalmente cognoscite le phenomenos de pression ossee super le medulla o le radice nervose o del presentia de un tense (curte) filo terminal, de un cyste, de un banda de constriction transverse, o de un tumor que pressa super le medulla.

Il esserea impossibile describer omne le anormalitates del medulla e del radices nervose spinal, proque hic como alterubi, il es quasi impossibile listar le defectos congenite. Il pare que le melior classification del anormalitates congenite del medulla spinal e del radices nervose spinal se basa super un separation del formas obvie ab le formas subtil. Esseva interprendite le tentativa de signalar certes del variationes subtil in le medulla e le radices nervose lumbar.

Le disveloppamento embryologic del medulla spinal determina su character bilatere. De accordo con iste facto, le varationes del medulla mesme es generalmente de forma bilatere. Il es estranie que in omne casos cognoscite de anomalia del radice nervose, le variation esseva de forma unilatere.

Le resection de un radice nervose non es un disastro major, sed illo deberea esser evitate, excepte quando on sape exactemente lo que on vole secar. Le implicationes del section de un radice nervose accessori differe ab le implicationes del section de un filo dichomatose.

Un inexplicate deformitate de pede con resistentia al correction o un anormal modo de ambulation sin traciabile factores etiologic deberea inducer le medico a prender in consideration le possibile presentia de anormalitates del medulla lumbar.

8

Dysplasia of the Neural Arch and Its Clinical Manifestation (Spondylolisthesis)*

EDUARD GÜNTZ AND KURT SCHLÜTER

It is the task of orthopaedics, and it has led to the name given this branch of medicine, to occupy itself with the straight upbringing of the child in the original meaning of the word. Hence, the congenital malformations of the skeletal apparatus have remained the undisputed domain of orthopaedics since the inception of that specialty. The frequency of congenital malformations differs between nations and races. One is reminded of the varying frequency of dislocation of the hip joint in whites and in Negroes. Certain congenital malformations of the skeletal apparatus are relatively common; for instance, clubfoot and dislocation of the hip. In these conditions medical experience has led to methods of treatment that are fairly uniform in the various medical centers. Thus, for example, there is unanimity as to the indications for earliest treatment of clubfoot, and also there is agreement in principle on the treatment of dislocated hip. It is certainly pleasant to have one's own views as to pathogenesis and therapy confirmed by other authors. However, it appears to the authors to be more fruitful to initiate a discussion of a morbid entity about which opinions still vary as to etiology, symptomatology and therapy.

We shall report on our experiences with spondylolisthesis. We know that opinions concerning this disease differ greatly even among German and other European authors. Therefore, we do not wish to neglect to mention in this chapter the views of others that one of us (Güntz) was able to acquire as an associate of Schmorl, as well as from the results of clinical and roentgenologic investigations made on patients seen at the Orthopaedic University Clinic "Friedrichsheim," Frankfort on the Main.

ON THE PATHOGENESIS OF SPONDYLOLISTHESIS

There is agreement in the Anglo-American and the German literature that the formation of a split in the interarticular portion is the prerequisite for the slipping of one vertebra on the vertebra below. We shall refrain from those forms of slipping of vertebrae that have been designated by Junghanns as pseudospondylolisthesis and are characterized by an altered position of the vertebral joints. We also wish to point out that recently in the German literature there has been discussion as to the inappropriate nomenclature (Brocher, Sonnenschein). However, we wish to retain the customary terminology so as not to complicate a critical comparison of the literature.

It may be advantageous to preface our exposition by a short ontogenetic consideration of the normal development of the vertebra. In the area occupied by the chorda dorsalis, cartilaginous nests are recognized

* From the Orthopaedic University Clinic "Friedrichsheim," Frankfort on the Main, Germany. Director: Professor E. Güntz. Translated from the German by Medical Literature Service, 19 S. 22nd St., Philadelphia 3, Pa.

during the 6th week of embryonal development. Later these nests take over the entire formation of the vertebral bodies. Dorsal to the vertebral body 2 processes form and constitute the neural arches. In the 4th embryonal month they unite behind the neural canal and complete the formation of the neural arch. An osseous nucleus appears as early as the 9th embryonal week in the vertebral body, and similar nuclei appear in the 11th embryonal week on both sides of the neural arch. Ossification proceeds only endochondrally. The neural arches also manifest perichondral ossification, such as is seen in long bones. Each process gives rise to one half of the neural arch, the pedicle and the articular processes, the transverse processes and one half of the spinal process. Between the 1st and the 2nd years of life, the 2 halves of the neural arch unite to form a vertebral arch. Between the 4th and the 6th years of life, the arch fuses with the vertebral body.

The interpretation of a split in the arch posteriorly as a manifestation of defective development does not offer any difficulties. Assuming the presence of only one center of ossification in each half of the neural arch, one cannot find an explanation of the split between the articular processes. Bardeen, Paturey, Keibel and Mall, Rambaud and Renault, and Willis have described 2 osseous nuclei for each half of the neural arch. However, new investigations by Bailey, Friberg, Raowe and Roche, and Tondury and Willis have shown that there is only one center of ossification in each half of the neural arch. Therefore, the assumption is untenable that one is dealing only with a simple inhibition of development.

Junghanns, referring to the spondylolyses encountered not infrequently in children (Brailsford, Capener, Eichlam, Garavano, George and Leonard, Guilleminet, Hitchcock, Jenkins, Johnstone and Thompson, Junghanns, Kleinberg, Kuttner, Langendorff, Meyerding, Mosberg, Priessnitz, Reisner, Rocher, Roederer, Schmorl, Silverskiold, Weil), still maintains that the formation of spondylolyses can be explained on a congenital, developmental basis. However, he also quotes the opinions of other authors who postulate that the splitting is caused by a scissorslike pressure action on the interarticular portion and resulting formation of an *Umbau* (rebuilding) zone (Lane, Mouchet and Roederer, Turner, Myer-Burgdorff).

An intermediate position is taken by Brocher, who has coined the concept of dysplasia of the vertebral arch on the basis of his roentgenologic observations and in analogy to the concept of hip joint dysplasia (Faber). According to him, the defective anlage of the arch is present even in the embryonal stage of development, but it becomes manifest only after the formation of the bony vertebra and the functional demands placed on the vertebral column. It is possible that during the growth period adaptations take place and prevent the appearance of an insufficiency. If during the period of puberty no further growth takes place, in-co-ordinated mechanical demands may produce a manifest disruption of continuity as the result of the dysplastic anlage.

Our clinical observations of the frequent occurrence of other malformations of the vertebrae associated with the presence of spondylolisthesis—for example, failure of the vertebral arches to unite posteriorly, dysplasia of the articular processes or hypoplasia of the slipping vertebra—point to the importance of disturbances in development, as does the increased incidence in the presence of numerical variations (Willis). Willis has pointed out that from the dorsal vertebral blood vessels the nutrient artery enters the vertebral arch at the interarticular portion. Therefore, it becomes clear that the presence of dysplasia results not only in mechanical but also in trophic growth disturbances. These may lead not only to a disruption in the continuity of the vertebral arch but also to a hypoplasia of the involved vertebra.

In our opinion, the increased familial incidence observed by Friberg and the hereditary transmission (Harbitz) would point also to the involvement of congenital malformation in the pathogenesis of spondylolisthesis. In our opinion also, the observations of Stewart on the increased incidence of spondylolisthesis in Eskimos living north of the Yukon support the concept of a hereditary etiology. This also has been cited by Brocher.

A 4th possibility for the genesis of spondylolisthesis—apart from the theories of a congenital etiology, a purely mechanical theory, and one due to dysplasia—is Hitchcock's theory of premature trophostatic etiology. The results of Hitchcock's investigations do not make it clear how a one-sided or a bilateral split formation in the arch of the 3rd lumbar vertebra could be possible in the absence of any changes in the 4th and the 5th lumbar vertebrae.

Finally, we should like to make note of the so-called traumatic spondylolisthesis (Albee, Ashbury, Boehler and Heuritsch, Darling, George and Leonard, Hadley, Holfelder, Lippens). It is not possible always to separate from the literature those observations in which a traumatic formation of the split is postulated by the authors—for example, the extremely rare observation of Bohler—and those in which a transition of a spondylolysis into a spondylolisthesis is meant.

Of course, it is true that many of our patients blame a traumatic event for their complaints, as will be discussed in more detail. Only in 2 cases were fractures observed, once of the transverse processes and once of the pelvis. After considering the mechanism of the trauma in these 2 cases, we believe that the split formation in the interarticular portion was not attributable to the trauma. Neither would we interpret Hadley's observations on fracture lines, new bone formation and callus in the spinal canal as the results of trauma; we feel that for the most part they represent spondylolyses and spondylolisthesis with a "traumatic history."

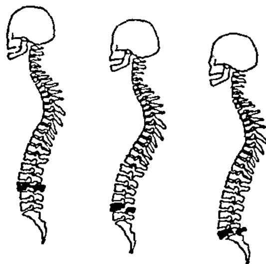
A closer analysis of the literature is beyond the scope of this chapter. In summary, we should like to confine ourselves to the assumption that the large majority of cases of spondylolisthesis are due to a congenital dysplasia of the vertebral arch, the manifestations of which appear relatively late. With a few exceptions in which a plausible traumatic mechanism is present, we are inclined to deny a traumatic etiology and to cite in support of this view the publications of Azema, Gerlach, Turner and Markellow.

The contrasting observations on the ossification of the vertebral arch will be the subject of further investigations by us. In this connection we should like to make some observations on the ossification of the tarsal bone. We found that with flatfeet the ossification of the calcaneus proceeded not infrequently from 2 endochondral centers. Similar observations were reported by Sever in the American literature. We conceive of this type of ossification process as an irregularity caused by altered physical conditions of the sphere of mechanical forces. We wish to leave open the question whether or not static moments in the ossification of the neural arch could be responsible for this abnormal course of ossification and explain the somewhat conflicting histologic findings.

Whereas congenital factors in the etiology of this entity appear to be more important to us than expositional factors, we consider a discussion of our observations under the chosen title to be justified. The therapeutic conclusions that we have drawn from our observations have permitted us to follow a method of treatment that differs in several particulars from the therapeutic measures of other authors. We should like to mention that we feel that operation is indicated in only a few instances.

OUTLINE OF PROBLEM

In the examination of our patients we observed that occasionally patients with a



L-3/4 = 3 Patients
= 2.41%

L-4/5 = 28 (7) Patients
= 22.59% (5.65%)

L-5/S-1 = 88 (31) Patients
= 70.98% (25.0%)

L-6/S-1 = 2 Patients
= 1.61%

L-3/4 + L-4/5 = 1 Patient
= 0.81%

L-4/5 + L-5/S-1 = 2 (1) Patients
= 1.61% (0.81%)

FIG. 1. Localization of 124 cases of spondylolisthesis seen over a 5-year period (1950-1955) at the Orthopedic University Clinic "Friedrichsheim," Frankfort on the Main. The figures in parentheses indicate the number of female patients or the percentage of female patients in the total patient population. The ratio of male to female patients is 85:39 (68.5%:31.5%).

high degree of slipping of a vertebra complained of minimal difficulties, whereas others with minimal slipping of a vertebra manifested severe functional disturbances. This observation, which certainly can be shared with the majority of orthopaedists, has prompted us to undertake a series of precise examinations in order to establish the clinically significant contribution of spondylolisthesis. Apart from intensive clinical and roentgenologic examinations of the static conditions, we have paid particular attention to muscle stretching and strain. Furthermore, we have examined patients for clinical signs of relaxation of the intervertebral disks and for disturbances in neurologic function.

In addition to observations recorded previously in the literature, we believe that we have made new observations deserving of reporting. On reading the literature, one easily may get the impression that the appearance of backaches and the roentgenologic demonstration of a "split" in the interarticular portion are sufficient to make a morbid entity. Such a concept certainly is incorrect. There is no lack of occasional hints of clinical symptoms, but apart from the investigations of Meyerding there are no large series of studies of the clinical symptomatology from the orthopaedic standpoint. The observations of others—for example, Ashbury, Azema, de Seze—may deal with a group of patients who showed symptoms different from those of our patients.

SURVEY OF OBSERVATIONS DURING THE YEARS 1950 TO 1955

A survey of 124 patients with spondylolisthesis observed and treated during the past 5 years is given in Figure 1. This confirms the statements of other authors that men are affected more often than women. Of our patients, 85 were men and 39 were women. Also, concerning localization we found no differences from the data given by other authors. More than two thirds of the observations concerned the slipping of the last lumbar vertebra over the sacrum. Next in frequency was the slipping of L-4 over L-5. In only 3 instances did we observe a spondylolisthesis at L-3 to L-4. We also observed 3 patients in whom 2 vertebrae were affected, in 1 case L-3/4 and L-4/5 and in 2 cases L-4/5 and L-5/S-1. The last-mentioned localization appears to be the rarest. It is not mentioned by Junghanns in the new edition of Schmorl-Junghanns,



FIG. 2. Male, born 1901, driver. Spondylolisthesis of L-3 over L-4 and L-4 over L-5. Roentgenogram taken in August, 1955. Complaints referable to the back first experienced 8 years before.



FIG. 3. Female, born 1938. Oblique roentgenogram taken in June, 1954, demonstrates the spondylolisthesis split in a case of spondylolisthesis involving L-4 over L-5 and L-5 over S-1. At L-4 the appearance of a Scotch terrier with a neck collar (Lachapele) is clearly recognizable.

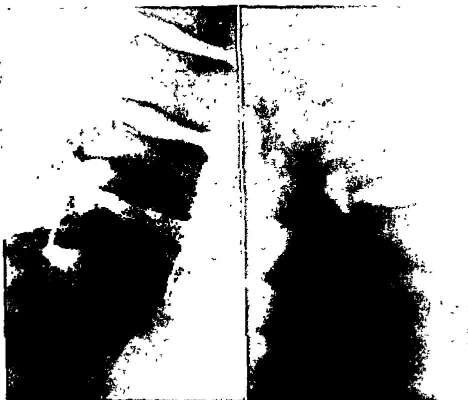


FIG. 4 Male, born 1911, foundryman. Roentgenograms of the spine in different positions show how on anteflexion (*left*) the intervertebral space between L-4 and L-5 narrows in its anterior portion, whereas on retroflexion (*right*) it widens anteriorly Clinically there was no evidence of damage of the disk.



FIG. 5. Male, born 1906, carpenter. Roentgenograms taken in different positions demonstrate that above the level of the spondylolisthesis at L-5/S-1 there is a distinct limitation of motion at L-4/L-5. Retroposition of L-4 over L-5. The functional deficit was related to damage of the disk between L-4 and L-5.

which carries an exhaustive bibliography. However, he cites the observation of Kovacs that a split formation was demonstrated in the interarticular portion of L-3, L-4 and L-5.

As an example of spondylolisthesis affecting 2 vertebrae, we should like to reproduce the roentgenograms of a 54-year-old patient. The lateral exposure shows clearly the slipping of L-3 over L-4 and of L-4 over L-5. Whereas marked changes are found in the disk between L-4 and L-5, the disk between L-3 and L-4 seems to have remained as yet essentially intact (Fig. 2).

The demonstration of the roentgenologic "split" is recognized by us also as the key to the diagnosis The best view of the split is in oblique exposures. Roentgenograms taken in the erect position from a distance are used by us to estimate slipping and to follow the course of the disorder in a given patient (Brocher, Dittmar, Jaeger, Glorieux, Güntz, de Seze) For the evaluation of the oblique exposures and for simple orientation, we would remind one of the little dog that Lachapele could recognize in the oblique picture of the vertebral arch and the vertebral processes. If the dog appears to have a neck collar, there is a break in the continuity in the bony part of the arch (Fig. 3). In Figure 3 one easily can recognize the formation of splits in 2 vertebral arches. Usually the demonstration of this cleft is successful with an oblique exposure when the split may be present in several planes when several vertebrae are affected. In our experience a degeneration of the intervertebral disk is not to be expected in all instances, as can be recognized from the motion pictures in a 44-year-old patient. With the patient bending forward and backward (Fig. 4), one can see clearly that the intervertebral space changes in its width. We have observed also that the loss of function cannot always be traced to a degeneration of the disk underlying the slipping vertebra, as is demonstrated in Figure 5. This is a 49-year-old patient in whom the loss of function was traced by clinical and roentgenologic means essentially to damage of the intervertebral disk at L-4 to L-5. This example shows that localization of the func-



FIG. 6. Female, born 1909, housewife. Follow-up roentgenograms taken in April, 1953 (*left*), and August, 1955 (*right*), show an increase in the degree of slipping of L-4 over L-5. The definite change in these findings was taken as evidence of increased slipping of L-4, although the 2 exposures were taken at slightly different projections.

tional disturbance is effected well by the taking of roentgenograms in various positions of the spine.

Our particular interest was devoted to the question of whether or not further slipping could be observed in the clinical course of the slipping of a vertebra. In the German literature the slipping of a vertebra is considered to be an indication for surgery (M. Lange). Actually, progressive slipping has rarely been observed, as pointed out especially by Brocher and Friberg. This fact has prompted Friberg to state that the fear of further slipping should not be interpreted as an indication for surgery.

We are able to communicate a few observations in which a progressive slipping and a progressive narrowing of the intervertebral space were noted. Examples are a 44-year-old female, shown in Figure 6, and



FIG. 7. Male, born 1919, excavation worker. In the follow-up roentgenograms taken in May, 1950 (*left*), and August, 1955 (*right*), one can recognize a progressive narrowing of the intervertebral space between L-5 and S-1 and an increase in the anterior displacement of L-5 over S-1.



FIG. 8. Male, born 1935, locksmith's apprentice. Follow-up roentgenograms of the lumbar spine taken in April, 1951 (*left*), July, 1953 (*center*), and August, 1955 (*right*), show the transition of a spondylolysis into spondylolisthesis with beginning anterior displacement of L-5 over S-1 (Grade 1 Meyerding). The straightening of the lumbar spine in the roentgenogram taken in 1951 is remarkable. In the last roentgenogram, taken in 1955, the position of the lumbar spine is normal. There are no significant symptoms.

a 31-year-old male, shown in Figure 7. Of 71 patients who were followed for 3 or more years, the observation of progressive slipping was certain in 3 cases and probable in 2. In 1 case, a 16-year-old youth, we were able to observe how a spondylolysis progressed to the 1st grade of spondylolisthesis. Roentgenograms taken from a distance (Fig. 8) show distinctly the changes in posture during the clinical course. Whereas the exposure taken in 1951 shows a stretched, straight posture of the lumbar spine as an expression of a protective posture (Güntz), the later roentgenograms reveal a progressive normalization of the position of the lumbar spine. The illustrations, and especially the roentgenograms taken in the various positions, illustrate that the separation and the slipping of the vertebra may occur before degeneration of the disk is recognizable roentgenologically.

It was shown also that a disturbance of function in the vertebral segment associated with motion is not present always by any means in the slipping vertebra. However, it is obvious that, in the presence of a degeneration of the disk, slipping of the vertebra may occur sooner. Even in the presence of a split in the interarticular portion, the slipping does not have to be in the anterior direction, as is demonstrated in Figure 9. This is a case of spondylolysis in a 29-year-old woman. One can see that L-4 is retropositioned over L-5. The direction of the displacement is determined by the position of the small vertebral joints, by the



FIG. 9. Female, born 1926, housewife. At L-4 there is a spondylolysis with typical split formation. Posterior displacement of L-4 over L-5 can be recognized.

plane of the articular surface and by the strength of the musculature and the ligamentous apparatus in addition to the strain pressure.

Retroposition of one vertebra over the next vertebra below in the presence of a damaged disk and with the prerequisite of a normal position of the joints and strong muscles with an intact ligamentous structure has been described by Güntz as a differential diagnostic criterion. Hirschberg and Brocher were able to demonstrate this phenomenon in experiments on the spinal column.

CLINICAL MANIFESTATIONS OF DEFICIT

We shall report on 71 patients who have been under treatment for prolonged periods

CLINICAL COURSE RELATED TO COMPLAINTS

NUMBER OF PATIENTS	PAINS EXPERIENCED ON:							PREFERRED POSITION AT WORK		SPORT: a) before b) after	
Site of Spondylolisthesis	Standing	Walking	Sitting	Lying Down	Up Steps	Down Steps	Lifting	Standing	Sitting	Motion	Making Diagnosis
14 Patients L-4/5											
Before Treatment	9	9	10	11	12	1	12	7	0	8	8
After Treatment	3	1	2	3	5	0	5	6	5	8	5
	3	6	5	3	3	1	5				
42 Patients L5/S1, L6/S1											
Before Treatment	27	21	29	31	28	8	30	11	6	29	28
	3	2	1	1	3	1	2			3	
After Treatment	7	4	5	2	5	6	8	9	12	34	6
	8	10	11	8	7	2	16	2	2	2	
3 Patients											
L3/4 before	1	1	1	3	3	0	3				
L3/4 +								2	0	2	2
L4/5 after		1	1	1	2	1	1	1	2	2	0
L4/5 + L5/S1	1			1			1				

The clinical course related to *individual complaints* in 59 patients treated conservatively. Figures set lower in the respective horizontal rows indicate that symptoms were experienced only occasionally or that work was possible only in certain body positions and with certain restrictions.

of time at the Clinic. Fifty-nine patients were treated conservatively, and 12 underwent surgery. The number of patients subjected to surgery is no greater among the total of 124 patients observed over the 5 years.

The table on page 79 summarizes the complaints of 59 patients treated conservatively. Although one deals here with the subjective complaints of patients who differ widely in their sensitivity to experience symptoms, nevertheless, in our opinion, valuable conclusions can be drawn from this summary. Patients were asked about pains on standing, walking, sitting, lying down, climbing steps and lifting objects. In addition, the patients expressed the position of the body that they favored in their work and in their sports activities. As was to be expected, most patients complained of pains when lifting objects. Almost of equal frequency were complaints of pains on climbing steps. Remarkably large was the number of patients who complained of pains on sitting. Few patients were able to pursue a sedentary oc-cupation. We should like to see in this [illegible] expression of the impossibility of a [illegible] kyphosis of the spinal column in the presence of maximally stretched muscles of the lumbar segment of the back. One deals [illegible] probably in only a few instances with a marked instability of the disk plate, the clinical picture of which has been described by Güntz as "loosening of the disk." [illegible] statement is supported by the rare occurrence of pain on descending steps, the "[illegible]ing pain" of the vertebral column. Nevertheless, complaints of this type were [illegible] common with lesions involving L-5/S-1. [illegible] was observed also by Brocher in 2 [illegible]. In such instances one deals with a [illegible] of the disk, i.e., a stage in which [illegible] slipping may be expected. An example [illegible] a female with a spondylolisthesis at L-[illegible] who complained of pains on descending steps and in whom further slipping [illegible] occurred (Fig. 6).

Not so common were complaints of [illegible]toms experienced on walking. [illegible] half of the patients with spondylolisthesis of L-5/S-1 voiced such complaints. [illegible] reason for this may be the possibility of [illegible]ternating, short-lasting relaxation of [illegible] strained muscles of the back.

The number of patients complaining [illegible] pains in the recumbent position [illegible]ably high. However, one has to consider [illegible] patients with kyphotic lumbar spine [illegible] lying on soft surfaces [illegible] ilar complaints to [illegible] on sitting. A [illegible]

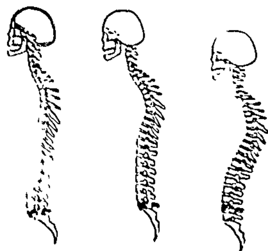


FIG. 10 Summarizes the types of spina[l] treated conservatively by the authors:

L-4/5	14 patients
L-5/S-1	39 patients
L-6 S-1	2 patients
Others	3 patients

The figures in parentheses refer to femal[e] spondylitis of the lumbar spine, was not includ[ed]

position includes patients with relaxation or loosening of the intervertebral disk. The largest group consists of those patients who have pressure pains in the recumbent position and clinically have percussion tenderness over the spinous process of the loosened part of the arch. This is an expression of the loosening of the arch demonstrable on operation. Patients protect their painful kyphosis in the presence of a loosened disk by placing a board under the mattress. They protect themselves from pressure pain over the spinous process of the loose arch by assuming the lateral position. This explains why eventually most patients have no complaints or minimal complaints when in the recumbent position, once the factors that bring about the pain have been elucidated and the patient has been given appropriate instructions.

During work it appears to be essential that the muscles be tensed and relaxed alternately. Therefore, most patients prefer an occupation in which they have an opportunity to move the body. Muscle exercises will have a favorable influence on the power to support the spinal column. Usually it would be a mistake to advise a sedentary job in the belief that such would relax the strain on the spine.

We have pondered whether or not root irritations could be the cause of the complaints in our patients, especially since the percentage of patients with neurologic symptoms has been reported to be very high by some authors (Ashbury in two thirds of his cases, Azema in 50% of cases, de Seze in over 60%). In our patients treated conservatively, only 6 out of 59 manifested neurologic symptoms. In this respect our observations are in agreement with those of Meyerding, who found manifestations of root compression in only 80 out of 745 cases. If we can find no grounds for attributing the complaints to root irritation, then we have to assume that muscular causes are responsible for the symptoms in the majority of cases. The cause of the pains would be an increased tonus of the musculature,



Fig. 11. Male, born 1910, construction worker. Spondylolisthesis at *L-5/S-1* associated with straightening of the lumbar and the thoracic spine. Onset of complaints 3 years before. Roentgenogram taken in August, 1955.

of time at the Clinic. Fifty-nine patients were treated conservatively, and 12 underwent surgery. The number of patients subjected to surgery is no greater among the total of 124 patients observed over the 5 years.

The table on page 79 summarizes the complaints of 59 patients treated conservatively. Although one deals here with the subjective complaints of patients who differ widely in their sensitivity to experience symptoms, nevertheless, in our opinion, valuable conclusions can be drawn from this summary. Patients were asked about pains on standing, walking, sitting, lying down, climbing steps and lifting objects. In addition, the patients expressed the position of the body that they favored in their work and in their sports activities. As was to be expected, most patients complained of pains when lifting objects. Almost of equal frequency were complaints of pains on climbing steps. Remarkably large was the number of patients who complained of pains on sitting. Few patients were able to pursue a sedentary occupation. We should like to see in this an expression of the impossibility of a further kyphosis of the spinal column in the presence of maximally stretched muscles of the lumbar segment of the back. One deals here probably in only a few instances with a marked instability of the disk plate, the clinical picture of which has been described by Güntz as "loosening of the disk." This statement is supported by the rare occurrence of pain on descending steps, the "poking pain" of the vertebral column. Nevertheless, complaints of this type were more common with lesions involving L-5/S-1, as was observed also by Brocher in 2 cases. In such instances one deals with a loosening of the disk, i.e., a stage in which further slipping may be expected. An example is a female with a spondylolisthesis at L-4/5 who complained of pains on descending steps and in whom further slipping actually occurred (Fig. 6).

Not so common were complaints of symptoms experienced on walking. Thus, only half of the patients with spondylolisthesis of L-5/S-1 voiced such complaints. The reason for this may be the possibility of alternating, short-lasting relaxation of the strained muscles of the back.

The number of patients complaining of pains in the recumbent position is remarkably high. However, one has to consider that patients with kyphotic lumbar spines when lying on soft surfaces ordinarily have similar complaints to those that they experience on sitting. A further group of patients who experience symptoms in the recumbent

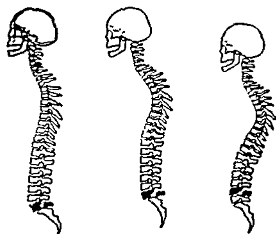


FIG. 10. Summarizes the types of spinal columns in spondylolisthesis in the patients treated conservatively by the authors:

	Type 1 (*Left*)	Type 2 (*Center*)	Type 3 (*Right*)
L-4/5 . . . . 14 patients	1 (1)	3	10
L-5/S-1 . . . . 39 patients	9 (2)	18 (2)	12 (3)
L-6/S-1 . . . . 2 patients	—	1	1
Others . . . . 3 patients	—	—	3

The figures in parentheses refer to females. 1 female, who also had a tuberculous spondylitis of the lumbar spine, was not included here.



FIG. 14. A 36-year-old patient with spondylolisthesis at L-5/S-1. Marked strain of the extensor muscles of the lumbar spine is clearly recognizable even in the resting position (*left*). Protective posture of the lumbar spine with marked fixation becomes distinctly evident on ventral flexion.

most commonly in juvenile patients at the onset of their complaints. The 2nd type consists of a stretched posture limited to the lumbar segment of the spine; it is encountered in older patients, especially in the subacute stage. It may be observed also in older patients in whom the thoracic spine permits only a limited extension. In such cases the entire upper portion of the body is tilted backward. In the 3rd type a significant alteration of the spinal curves is not recognizable. This type is observed with fibrous stiffening of the segment involved in motion. Roentgenologically one finds a narrowing of the intervertebral space and reactive changes in the form of console-like excrescences. In this group we see spondylolisthesis as an accidental finding without clinical significance. The different posture types can be demonstrated well by means of roentgenograms taken from a distance.

In order to demonstrate also the postural changes of the thoracic segments, we have mounted the roentgenograms of the thoracic spine taken in the erect position over the lumbar spine. Figures 11 to 13 illustrate the 3 postural types as they appear in roentgenograms. The same changes are expressed also in the clinical picture, although here the strained muscles may protrude so far over the spinous processes that the examiner may fail to notice a flattening of the lumbar curve. But always we can observe the fixation of the spine on motion as illustrated in Figures 14 and 15. The most severe muscle strain can be observed at the onset of the complaints. Patients with strong muscles permit the recognition of a saddle-shaped flattening of the lumbar spine sooner than do patients with weak muscles. In the latter, kyphosis of the lumbar spine on straightening of the pelvis may remain even after the end of the acute stage.

We believe that we are justified in concluding from our observations that the backaches and the "sciatic pains" in the majority of patients are caused by muscle strain. In this connection, apart from strain of the back muscles, we found also straining of the ischiocrural musculature in 16 cases. The most severe disturbances of posture are found in those who have a loosening of the intervertebral disk. This observation often is made with only minimal slipping corresponding to Meyerding's Grade 1. In this connection we should like to confirm the observations of Glorieux and Roederer that the onset of complaints coincides with the



FIG. 12 (*Left*). Male, born 1910, locksmith. Spondylolisthesis at L-5/S-1 associated with straightening of the lumbar spine and wide, arched lumbar scoliosis. Frequently such scolioses show no compensatory curves at the thoracic level. In this case the thoracic spine manifested a normal degree of kyphosis. Roentgenogram taken in August, 1955. Onset of complaints 12 years before.

FIG. 13 (*Bottom*). Male, born 1919, potter. Roentgenogram taken in August, 1955. Spondylolisthesis at L-4/L-5 with normal curves of the spinal column in the sagittal plane. 11 years previously the patient was in an accident, in which he suffered compression trauma of the back and tear fractures of the right transverse processes between L-2 and L-4, since which time he had suffered backaches.



which is tensed maximally in order to secure a protective posture. Stretched positions of the spine are clearly recognizable in the roentgenograms.

Three types of posture could be differentiated; they are summarized in Figure 10. The 1st type, which consists of a stretched posture of the entire spine, is encountered

Burckhardt, H.: Arch. klin. Chir. 157:69, 1929.
———: Deutsche Ztschr. Chir. 232:25, 1931.
Charry, V.: Rev. orthop. 23:245, 1936.
Cleveland, M., Bosworth, D., and Thompson, F. R.: J. Bone & Joint Surg. 30A:302, 1948.
da Silva, Martins: J. belge radiol. 35:284-303, 1952.
De Sèze: Quoted by Brocher.
De Sèze and Dijan: Rev. rhum. 18:111, 1951.
De Sèze and Durieux: Semaine hôp. Paris 26: 401, 1950.
Diessl, F.: Zorg. Chir. 51:264, 1929.
Ehrlich: Arch. orthop. 32:638, 1933.
Eichlam, K.: Zentralbl. Chir. 61:555-558, 1934.
Epstein, G.: Fortschr. Geb. Röntgenstrahlen 46:344, 1932.
Faber, A.: Untersuchung über die Ätiologie und Pathologie der angeborenen Hüftverrenkungen, Leipzig, Thieme, 1938.
Faldini, G.: Zentralbl. Chir. 56:3115, 1929.
Follmer, B.: Med. Monatsschr. 1:366, 1947.
Francillon, M. R.: Die Medizinische, p. 451, 1953.
———: Schweiz. med. Wchnschr. 80:1256, 1950.
Friberg, Sten.: Studies on spondylolisthesis, Acta chir. scandinav. 82:55, 1939.
Friedl, E.: Röntgenpraxis 7:374-379, 1935.
Gajzago: Monatsschr. Geburtsh. 95:54, 1933.
Garavano, P.: Zorg. Chir. 52:107, 1931.
Gaugele, K.: München. med. Wchnschr. 77: 1992, 1930.
George and Leonhard: The vertebrae, roentgenologically considered, Ann. Roentgenol vol. 8, 1929.
Gerlach, G.: Arch. orthop. u. Unfall.-Chir. 33: 464, 1933.
———: Ztschr. orthop. Chir. 58:465, 1933.
Gillespie, H : Brit. J. Radiol. 24:193-197, 1951.
Gjessing, Morten. H.: Acta orthop. scandinav. 20:200-213, 1951.
Glorieux, P., and Roederer, C.: La spondylolyse et ses conséquences, Paris, Masson, 1937.
Grashey, R.: Röntgenpraxis 5:387, 1933.
———: Röntgenpraxis 5:388, 1933.
———: Röntgenpraxis 6:197, 1934.
Guerrini, G., and Ghislanzoni, R.: Radiologica (Roma) 8:287-307, 1952.
Guilleminet, M.: Rev. orthop. 23:385, 1936.
———. Zorg. Chir. 76:566, 1936.
Guilleminet, M., and Lacour: Presse méd. 59:101-102, 1951.
Güntz, E.: Arch orthop. u. Unfall-Chir. 34: 333-355, 1934.
———: Röntgenpraxis 6:224-228, 1934.
———: Röntgenpraxis 8:73-87, 1937.
———: Verhandl. deutsch. orthop. Gesellsch., 31st Congress, Königsberg, August 28-30, 1936, pp. 245-257, Stuttgart, Enke, 1937.
———: Schmerzen und Leistungsstörungen bei Erkrankungen der Wirbelsäule, Stuttgart, Enke, 1937.
———: Deutsche Ztschr. Chir. 254:633-648, 1941.
———: Schäden der Wirbelsäule *in* Fischer, Herget, and Molineus: Das ärztliche Gutachten im Versicherungswesen, München, Barth, 1955.
Hadley, L. A.: J. Bone & Joint Surg. 37A:787, 1955.
Hammerbeck, W.: Fortschr. Geb. Röntgenstrahlen 54:144, 1936.
Harbitz, H.: Zorg. Chir. 73:192, 1935.
Harris, R. I.: Spondylolisthesis, Ann. Roy. Coll. Surgeons England 8:259-297, 1951.
———: J. Bone & Joint Surg. 37A:786, 1955.
Harttung, H.: Beitr. klin. Chir. 150:269, 1930.
Hasselwander, A.: Handbuch der Anatomie des Kindes, München, Bergmann, 1938.
Hayek, H.: Zentralbl. Gynäk. 52:2511, 1928.
Hellner, H.: Fortschr. Geb. Röntgenstrahlen 41:527, 1930.
Hepp, O.: Arch. orthop. u. Unfall-Chir. 39: 728, 1939.
Hirsch, M.: Zentralbl. Chir. 57:1733, 1930.
Hitchcock: Quoted by Brocher.
Hohmann, G.: Ztschr. ärztl. Fortbild. 35:616, 1938.
Holfelder, H.: Röntgenpraxis 2:865-875, 1930.
Huber, E.: Monatsschr. Unfallh. 38:301, 1931.
Hulbert, N. G.: Proc. Roy. Soc. Med. 41:97, 1948.
Iles, A.: Brit. J. Radiol. 8:659, 1935.
Ingerbrigtsen, R.: Acta chir. scandinav. 105: 172-181, 1953.
Janssen, K.: Röntgenpraxis 5:742, 1933.
Jenkins, J.: Brit. J. Surg. 24:80, 1936.
Johnson, R.: J. Bone & Joint Surg. 16:867, 1934.
Johnstone and Thompson: Zentralbl. Radiol. 19:620, 1935.
Joisten, Chr.: Zentralbl. Chir. 56:2312, 1929.
Jonkhere and Leclerq: Zorg. Chir. 78:821, 1936.
Junghanns, H.: Arch. klin. Chir. 159:423, 1930.
———: Beitr. klin. Chir. 148:554, 1930.
———: Fortschr. Geb. Röntgenstrahlen 41: 239, 1930.
Keller, E.: Chirurg 9:216, 1937.
Klar: Verhandl. deutsch. Röntgen-Ges. 23:50, 1931.

SUMMARY

After a short survey of normal ossification of the vertebral arch, the various theories of split formation in the interarticular portion were discussed. The authors are of the opinion that dysplasia of the vertebral arch (Brocher) is a prerequisite of the formation of the split.

The authors report on 124 cases of spondylolisthesis, including 3 cases involving the slipping of 2 neighboring vertebrae. Seventy-one patients were available for prolonged observation and critical re-evaluation.

Since symptoms of neurologic deficit were demonstrable in only 6 instances, the authors consider it improbable that root irritation is responsible for the symptoms complained of by the majority of patients with spondylolisthesis. Numerous patients manifested defective postures of the spine either confined to the lumbar portion or involving the entire spinal column. Such protective postures have been described as clinical symptoms of relaxation of the intervertebral disk (Güntz). In some patients it was possible actually to demonstrate clinically the presence of disk relaxation. In these patients the possibility of progressive vertebral slipping was assumed and could be demonstrated actually by roentgenologic means. Apparently the onset of clinical complaints coincides with the onset of slipping. In the majority of cases the complaints were characterized by muscle strain that extended up to the thoracic part of the back muscles and down to the ischiocrural muscles (pseudosciatica).

In spondylolisthesis a mechanical insufficiency of the passive motor apparatus leads to a straining of the musculature. The strength of the musculature and of the ligamentous apparatus, the condition of the disk, and the mechanical properties of the inclination of the slipping vertebra and of the slipping surface determine the course of the slipping. Before embarking on treatment it is necessary to establish what causes symptoms. In treating spondylolisthesis, one cannot treat a single entity, instead, one has to treat various disturbances of the motor apparatus of the spine—for example, relaxation of the disk, muscle strain and, in rare instances, root irritation. By means of an exact analysis, aimed therapy becomes possible.

Of 124 patients, 12 were treated surgically. The indications for surgery are as follows:

1. The lack of significant improvement after a year of conservative therapy (also after the wearing of a support)
2. Recurrent sciatic root pain in the presence of spondylolisthesis
3. Demonstrable progressive slipping of vertebra in the presence of disk relaxation

BIBLIOGRAPHY

Abraham, H.: Über Wirbelgleiten (Spondylolisthese), unter Zugrundelegung von 23 Beobachtungen der Rontgenabteilung der Chirurgischen Universitätsklinik zu Frankfurt a. M. Diss. Frankfurt a. M. 1934.

Ach: Arch. klin. Chir. 163:640, 1931.

Adkins, E. W. O.: J. Bone & Joint Surg. 37B: 208, 1955.

Arden, G. P.: Proc. Roy. Soc. Med. 42:601, 1949.

Ashbury: Quoted by Brocher.

Azema, M. A.: Le spondylolisthesis, thesis, Paris, Jouve, 1932.

Bailey, W.: Radiology 48:107, 1947.

Bardeen: Quoted by Th. Willis

Batts, M.: The etiology of spondylolisthesis, J. Bone & Joint Surg. 22:879, 1939.

Benassi, G.: Zentralbl. Radiol. 20:104, 1935.

Bickel, W. H.: J. Bone & Joint Surg. 37A:786, 1955.

Blume, W.: Ztschr. Anat. u. Entwcklngsgesch. 101:719-745, 1933.

Bohler, L.: Chirurg 7:477, 1935.

Böhler and Heuritsch: Chirurg 6:485, 1934.

Bosworth, D. M., Fielding, J. W., deMarest, L., and Bonaquist, M.: J. Bone & Joint Surg. 37A:767, 1955.

Brailsford, J.: Brit. J. Radiol 6:666, 1933.

Briggs, H., and Keats, S.: J. Bone & Joint Surg. 29:328, 1947.

Brocher, J. E. W.: Fortschr. Geb. Röntgenstrahlen 73:719-726, 1950.

———: Schweiz. med. Wchnschr. 83:788, 1953.

———: Die Wirbelverschiebung in der Lendengegend, Leipzig, Thieme, 1951.

Burckhardt, H.: Arch. klin. Chir. 157:69, 1929.
———: Deutsche Ztschr. Chir. 232:25, 1931.
Charry, V.: Rev. orthop. 23:245, 1936.
Cleveland, M., Bosworth, D., and Thompson, F. R.: J. Bone & Joint Surg. 30A:302, 1948.
da Silva, Martins: J. belge radiol. 35:284-303, 1952.
De Sèze: Quoted by Brocher.
De Sèze and Dijan: Rev. rhum. 18:111, 1951.
De Sèze and Durieux: Semaine hôp. Paris 26: 401, 1950.
Diessl, F.: Zorg. Chir. 51:264, 1929.
Ehrlich: Arch. orthop. 32:638, 1933.
Eichlam, K.: Zentralbl. Chir. 61:555-558, 1934.
Epstein, G.: Fortschr. Geb. Röntgenstrahlen 46:344, 1932.
Faber, A.: Untersuchung über die Ätiologie und Pathologie der angeborenen Hüftverrenkungen, Leipzig, Thieme, 1938.
Faldini, G.: Zentralbl. Chir. 56:3115, 1929.
Follmer, B.: Med. Monatsschr. 1:366, 1947.
Francillon, M. R.: Die Medizinische, p. 451, 1953.
———: Schweiz. med. Wchnschr. 80:1256, 1950.
Friberg, Sten.: Studies on spondylolisthesis, Acta chir. scandinav. 82:55, 1939.
Friedl, E.: Röntgenpraxis 7:374-379, 1935.
Gajzago: Monatsschr. Geburtsh. 95:54, 1933.
Garavano, P.: Zorg. Chir. 52:107, 1931.
Gaugele, K.: München. med. Wchnschr. 77: 1992, 1930.
George and Leonhard: The vertebrae, roentgenologically considered, Ann. Roentgenol. vol. 8, 1929.
Gerlach, G.: Arch. orthop. u. Unfall.-Chir. 33: 464, 1933.
———: Ztschr. orthop. Chir. 58:465, 1933.
Gillespie, H.: Brit. J. Radiol. 24:193-197, 1951.
Gjessing, Morten. H.: Acta orthop. scandinav. 20:200-213, 1951.
Glorieux, P., and Roederer, C.: La spondylolyse et ses conséquences, Paris, Masson, 1937.
Grashey, R.: Röntgenpraxis 5:387, 1933.
———: Röntgenpraxis 5:388, 1933.
———: Röntgenpraxis 6:197, 1934.
Guerrini, G., and Ghislanzoni, R.: Radiologica (Roma) 8:287-307, 1952.
Guilleminet, M.: Rev. orthop. 23:385, 1936.
———: Zorg. Chir. 76:566, 1936.
Guilleminet, M., and Lacour: Presse méd. 59:101-102, 1951.
Güntz, E.: Arch orthop. u. Unfall-Chir. 34: 333-355, 1934.
———: Röntgenpraxis 6:224-228, 1934.
———: Röntgenpraxis 8:73-87, 1937.
———: Verhandl. deutsch. orthop. Gesellsch., 31st Congress, Königsberg, August 28-30, 1936, pp. 245-257, Stuttgart, Enke, 1937.
———: Schmerzen und Leistungsstörungen bei Erkrankungen der Wirbelsäule, Stuttgart, Enke, 1937.
———: Deutsche Ztschr. Chir. 254:633-648, 1941.
———: Schäden der Wirbelsäule *in* Fischer, Herget, and Molineus: Das ärztliche Gutachten im Versicherungswesen, München, Barth, 1955.
Hadley, L. A.: J. Bone & Joint Surg. 37A:787, 1955.
Hammerbeck, W.: Fortschr. Geb. Röntgenstrahlen 54:144, 1936.
Harbitz, H.: Zorg. Chir. 73:192, 1935.
Harris, R. I.: Spondylolisthesis, Ann. Roy. Coll. Surgeons England 8:259-297, 1951.
———: J. Bone & Joint Surg. 37A:786, 1955.
Harttung, H.: Beitr. klin. Chir. 150:269, 1930.
Hasselwander, A.: Handbuch der Anatomie des Kindes, München, Bergmann, 1938.
Hayek, H.: Zentralbl. Gynäk. 52:2511, 1928.
Hellner, H.: Fortschr. Geb. Röntgenstrahlen 41:527, 1930.
Hepp, O.: Arch. orthop. u. Unfall.-Chir. 39: 728, 1939.
Hirsch, M.: Zentralbl. Chir. 57:1733, 1930.
Hitchcock: Quoted by Brocher.
Hohmann, G.: Ztschr. ärztl. Fortbild. 35:616, 1938.
Holfelder, H.: Röntgenpraxis 2:865-875, 1930.
Huber, E.: Monatsschr. Unfallh. 38:301, 1931.
Hulbert, N. G.: Proc. Roy. Soc. Med. 41:97, 1948.
Iles, A.: Brit. J. Radiol. 8:659, 1935.
Ingerbrigtsen, R.: Acta chir. scandinav. 105: 172-181, 1953.
Janssen, K.: Röntgenpraxis 5:742, 1933.
Jenkins, J.: Brit. J. Surg. 24:80, 1936.
Johnson, R.: J. Bone & Joint Surg. 16:867, 1934.
Johnstone and Thompson: Zentralbl. Radiol. 19:620, 1935.
Joisten, Chr.: Zentralbl. Chir. 56:2312, 1929.
Jonkhere and Leclerq: Zorg. Chir. 78:821, 1936.
Junghanns, H.: Arch. klin. Chir. 159:423, 1930.
———: Beitr. klin. Chir. 148:554, 1930.
———: Fortschr. Geb. Röntgenstrahlen 41: 239, 1930.
Keller, E.: Chirurg 9:216, 1937.
Klar: Verhandl. deutsch. Röntgen-Ges. 23:50, 1931.

Kleinberg, S.: J. Bone & Joint Surg. 16:441, 1934.
Kleinhaus, E.: Fortschr. Geb. Röntgenstrahlen 37:335, 1928.
Kopitz, J.: Arch. orthop. u. Unfall.-Chir. 34: 609, 1934.
Kovacs, A.: Acta Medica 2:427-430, 1951.
Küttner, H.: Zentralbl. Chir. 54:404, 1927.
Lachapèle, A.: Bull. soc. electroradiol. France 27:175, 1939.
Lane and Moore: Ann. Surg. 127:537, 1948.
Lang, F.: Monatsschr. Unfallh. 43:566, 1936.
Lange, M.: Orthop. Chir. Operationslehre, München, Bergmann, 1951.
Langendorff, G.: Ztschr. Orthop. 83:548-557, 1953.
Lawson, J.: J. Bone & Joint Surg. 14:387, 1932.
Le Double: *Traité des variations de la colonne* vertébrale de l'homme et de leur signification au point de vue de l'anthropologie zoologique, Paris, 1912.
Liechti, A.: Die Röntgendiagnostik der Wirbelsäule und ihre Grundlagen, ed. 2, Wien, Spinger, 1948.
Lindemann, K., and Kuhlendahl, H.: Die Erkrankungen der Wirbelsäule, Stuttgart, Enke, 1953.
Lippens, A.: Presse méd. 42:622-624, 1934.
———: *Rev. Physiol. et Pathol., p. 155, 1932.*
Litten, F.: Röntgenpraxis 4:1039, 1932.
MacNab, I.: J. Bone & Joint Surg. 32B:325, 1950.
Marique, Laurent, van Gaver and van Hove: Acta chir. belg., supp. 3, 1951.
Mathieu and Demirleau: Rev. orthop. 23: 352, 1936.
Mercer, W.: Zorg. Chir. 80:591, 1936.
Meyer, H.: Arch. orthop. u Unfall.-Chir. 29: 109, 1930.
———: Zentralbl Chir. 57:2619, 1930.
Meyer-Burgdorff, H.: Arch. orthop. u. Unfall.-Chir. 29:109, 1930.
———: Beitr. klin. Chir. 151:386, 1931.
———: Deutsche Ztschr. Chir. 245:173, 1935.
———: Zentralbl. Chir. 58:1137, 1931.
Meyer-Burgdorff, H., and Klose-Gerlich, J.: Arch. klin *Chir. 182:220, 1935.*
Meyerding, H.: J. Bone & Joint Surg. 23:461, 1941.
———: Monatsschr. Unfallh. 38:334, 1931.
———: Radiology *20:108, 1933.*
Michalowski, B.: Zentralbl. Chir. 51:2360, 1927.
Mjaktotnich, S.. Zorg Chir. 60:615, 1933.
Mosberg, G.: Zentralbl Radiol. 20:256, *1935.*
Mouchet, A.: Rev. orthop. 22:97, 1935.
Mouchet, A., and Roederer, C.: Presse méd. 39:569, 1931.
———: Rev. orthop. 13:461, 1927.
Müller, W., and Zwerg, H.: Beitr. klin. Chir. 149:155, 1929.
Neugebauer, F.: Arch. Gynäk. 20:133, 1882.
Nissen-Lie, H. S.: Nord. med. 11:431, 1941.
Novak, J.: Zentralbl. Gynäk. *52:2502, 1928.*
Orth, O.: Röntgenpraxis 8:206, 1936.
Pannhorst, R.: Deutsche med. Wchnschr., p. 506, 1942.
Paturet, G.: Traité d'anatomie humaine, vol. 1, Paris, Masson, 1951.
Pavlik, A.: Zorg. Chir. 79:192, 1936.
Perlman, R., and Hawes, L.: J. *Bone & Joint* Surg. 33A:1012-1013, 1951.
Picot, M.: Zentralbl. Neur. 49:67, 1928.
Priessnitz, O.: Med. Klinik 45:500, 1950.
Rambaud, A., and Renault, Ch.: Origine et développement des os, Paris, F. Chamerot, 1864.
Rathcke, L.: Deutsche med. Wchnschr. 63: 1228, 1937.
———: Arch. orthop. u. Unfall-Chir. 43:111, 1944.
Reinbold, P.: Schweiz. Ztschr. Unfallmed. 25: 197, 1931.
———: Beitr. klin. Chir. 162:64, 1935.
Reischauer, F.: Fortschr. Geb. Röntgenstrahlen 58:343, 1938.
Reisner, A.: Arch. orthop. u. Unfall-Chir. 32: 135, 1932.
Rocher, H.: Zentralbl. Chir. 58:420, 1931.
Rocher, H., and Roudil: Zorg. Chir. 47:694, 1929.
Roederer, C., and Glorieux, P.: Presse méd. *41:1550, 1933.*
Rowe, G. G., and Roche, M. B.: J. Bone & Joint Surg. 35A:102, 1953.
Ruhnau, A.: Über Spondylolisthesis, Diss. Königsberg, 1932.
Ryden, A.: Zentralbl. Chir. 58:428, 1931.
Ryzow, J.: Zorg. Chir. 58:543, 1932.
Schaer, H.: Beitr. klin. Chir. 155:287, 1932.
Schanz, A.: Die Lehre von den statischen Insuffizienzerkrankungen mit besonderer Berücksichtigung der Insufficienta vertebrae, Stuttgart, Enke, 1921.
Scherb, R.: Ztschr. orthop. Chir. 50:304, 1928.
Schluter, K.: Beitr. klin. Chir. 191:257-268, 1955.
———: Verhandl. deutsch. orthop Gesellsch., 43rd Congress, Hamburg, September 14-17, 1955, pp. 335-343, Stuttgart, Enke, 1956.
———: *Ztschr. orthop. Chir. 187:37-55,* 1955.

Schmorl, G.: Deutsch. Ztschr. Chir. 237:422, 1932.
Schmorl, G., and Junghanns, H.: Die gesunde und die kranke Wirbelsäule in Röntgenbild und Klinik, ed. 3, Stuttgart, Thieme, 1953.
Schüller, J.: Med. Monatsschr. 3:529, 1949.
———: Zentralbl. Chir. 58:1085, 1931.
Schulz, A.: Ztschr. Orthop. Chir. 49:546, 1928.
Schwegel: Quoted by Junghanns.
Sever, J. W.: Surg., Gynec. & Obst. 50:1012, 1930.
Sicard, A., and Leca, A.: Presse méd. 60:914-918, 1952.
Silverskiöld, N.: Acta orthop. scandinav. 1:42, 1930.
Sisefsky, M.: Acta orthop. scandinav. 4:234, 1933.
Smirnow, A.: Zentralbl. Chir. 54:328, 1927.
Sonnenschein, A.: Arch. orthop. u. Unfall-Chir. 46:588, 1954.
Sprung, H.: Deutsch. Ztschr. Chir. 249:632, 1938.
Stepin, S.: Zorg. Chir. 50:369, 1930.
Stewart, J.: Zorg. Chir. 55:597, 1931.
Stewart, T.: J. Bone & Joint Surg. 17:640, 1935.
Suermont, W.: Zorg. Chir. 57:245, 1932.
Tamini, L.: Zentralbl. Chir. 56:378, 1929.
———: Rev. chir. 39:553, 1953.
———: Biol. Berichte 92:156, 1955.
Töndury, G.: Ztschr. Anat. 110:568, 1940.
Turner, H.: Ztschr. orthop. Chir. 51:23, 1929.
Turner, H., and Markellow: Acta chir. scandinav. 67:914, 1930.
Turney, J. P.: Brit. M. J. 11:128, 1952.
Waard, T.: Zorg. Chir. 54:697, 1931.
Wallgren, G.: Acta orthop. scandinav. 4:23-43, 1933.
Wegener, E.: Arch. orthop. u. Unfall-Chir. 26:73, 1928.
———: München. med. Wchnschr. 74:299, 1927.
Weil, P.: München. med. Wchnschr. 78:387, 1931.
———: Zentralbl. Chir. 54:1251, 1927.
Weil, S.: Zentralbl. Chir. 57:2620, 1930.
Wiemers, A.: Arch. orthop. u. Unfall-Chir. 12:387, 1913.
Wilhelm, R.: Arch. orthop. u. Unfall-Chir. 24:189, 1927.
Willis, Th.: Am. J. Anat. 32:95, 1924.
———: J. Bone & Joint Surg. 13:709, 1931.
———: J. Bone & Joint Surg. 14:267, 1932.
Wolff, G.: Ztschr. orthop. Chir. 63:133, 1935.
Wunderlich, H.: Arzt und Sport 12:1, 1936.
Wyss, A.: Schweiz. med. Wchnschr. 80:1257-1258, 1950.
Zwerg, H.: Verhandl. deutsch. Röntgen Gesellsch. 23:44, 1931.

Dysplasia del Arco Neural e su Manifestation Clinic (Spondylolisthese)

Eduard Güntz e Kurt Schlüter

Summario in Interlingua

Post un breve revista del normal ossification del arco vertebral, le varie theorias del formation findite in le portion interarticular es discutite. Le autores opina que dysplasia del arco vertebral (Brocher) es un precondition pro le formation del fissura.

Le autores reporta 124 observationes de spondylolisthese, includente 3 casos de displaciamento de duo vertebras adjacente. Septanta-un patientes esseva dispondibile pro observationes prolongate e re-evaluationes critic.

Proque symptomas de deficit neurologic esseva demonstrabile in solmente sex casos, le autores considera como improbabile que irritation de radice esseva responsabile pro le symptomas del quales se plangeva le majoritate del patientes con spondylolisthese. Numerose patientes manifestava defective posturas del spina dorsal in tanto que le erectura esseva restringite al portion lumbar del spina dorsal o involveva le spina integre. Tal posturas protective ha essite describite como symptomas clinic del relaxation del disco intervertebral (Güntz). In un certe numero de patientes il esseva de facto possibile demonstrar clinicamente le presentia de relaxation de disco. In iste patientes le possibilitate de progressive displaciamento vertebral esseva stipulate e se provava de facto demonstrabile per medios radiologic. Le declaration de disturbationes clinic coincide apparentemente con le declaration del displaciamento. In le majoritate del casos le

disturbationes esseva characterisate per un tension muscular que se extendeva in alto usque al parte thoracic del musculos dorsal e in basso usque al musculos ischio-crural (pseudo-sciatica).

In spondylolisthese un insufficientia del passive apparato motor resulta in un tension del musculatura. Le fortia del musculatura e del apparato ligamentose, le condition del disco, e le qualitates mechanic del vertebra e del superficie involvite in le displaciamento es le factores que determina le curso del displaciamento. Ante que on initia le therapia il es necessari establir le causas del symptomas. Quando on tracta spondylolisthese on non pote tractar un entitate isolate. In loco de isto, on debe tractar varie disturbationes del apparato motor del spina vertebral, per exemplo le relaxation de discos, tensiones muscular, e in rar casos le irritation de radices. Per medio de un exacte analyse on pote elaborar un therapia a orientation planate.

Ex le total de 124 patientes, 12 esseva tractate chirurgicamente. Le indicationes pro intervention chirurgic es le sequente:

1. Le absentia de significative meliorationes post un anno de tractamento conservative (includente le uso de un apparato de contention).

2. Recurrente dolores sciatic de radice in le presentia de spondylolisthese.

3. Demonstrabilitate del displaciamento del vertebra in le presentia de relaxation de disco.

9

Splinting for Controlled Movement

Denis Browne, F.R.C.S.*

PREVIEW

In this chapter it is suggested that certain congenital deformities are caused by mechanical forces acting on the fetus in utero, and that from this hypothesis of causation valuable principles in treatment may be deduced. These principles are (1) that similar forces to those assumed to have caused the deformity should be used to reduce it and (2) that it is essential to allow active movement during the process of correction in order to counter the bad effects of immobilization and displacement of the parts concerned before birth.

In order to observe these principles, various original splints and devices are described, together with the common variations and modifications of them. These modifications are criticized, and reasons for not considering them to be improvements are given.

The conditions described are:

1. The 3 standard varieties of molding of the feet: clubfoot; metatarsal varus; metatarsal valgus.
2. Congenital dislocation of the hip
3. Congenital postural scoliosis, which is argued to be the early stage of "idiopathic scoliosis"
4. Sternocleidomastoid torticollis

DEFINITION OF SPLINTING

Splinting may be defined as the control of movement of bones by artificial means. It can be of 2 kinds: the first is immobilization to eliminate all movement; the second is controlled movement, in which certain movements are prevented as harmful and others are permitted as beneficial. For certain conditions, such as inflammations and fractures, complete stillness is needed during healing, but I would suggest that there are other types of human ills for which the best treatment includes the encouragement of growth and the development of muscle balance given by controlled movement.

The method of treatment outlined here is based upon two of the teachings of Hippocrates: the first is that certain deformities, such as talipes, are due to mechanical molding in the uterus before birth; the second is that active use encourages growth and that its lack causes atrophy.

I have stated elsewhere[1] my reasons for accepting the first assumption, and I know of no reasonable attempt to controvert them by anyone who has studied what they actually are. The difficulty about getting them accepted generally is that they depend not upon experiment, that is, deliberate altering of the course of nature, but upon observation and reasoning. To take classic examples, the difference between the experimental method and the observational one is the difference between Pasteur and Harvey on the one side and Newton and Einstein on the other. It seems to me a pity that at present medicine depends almost entirely upon experiments, influenced by the brilliant results gained in this way in the past.

These mechanical inter-uterine forces may

*Surgeon to the Hospital for Sick Children, Great Ormond Street, London

have 4 effects, the consideration of which gives useful clues to treatment.

MOLDING

If one assumes that parts of the body can be molded before birth in the way that it is admitted universally that they can be molded afterward (in such instances as the feet of Chinese ladies) we are led toward the assumption that the best way to correct these moldings would be to use similar forces in the reverse direction.

The characteristics of these forces are:

1. *They are persistent* and so differ completely from manipulations in which a contracted part may be stretched but on the cessation of the manipulation is allowed to contract again.

2. *They are varying or pulsating,* changing in force with the movements of the fetus, and so differ from the unchanging pressure given by a plaster cast. This is most important, as such pulsations pump the blood through the tissues, so avoiding that stagnation that is one of the main factors in pressure sores. In this way pressures far greater than those permissible by means of immobilization may be used safely.

3. *They are distributed widely by soft surfaces,* in particular the uterine walls, which yield over bony points and so avoid pressure necrosis in these sites.

4. *They produce no friction,* owing to the lubricating effect of the fluid surrounding the fetus.

DISPLACEMENT

This is obvious enough in such conditions as congenital dislocation of the hip, due to a thrust on the knee, or dislocation of the knee due to the leg's being in a wrong position with the foot up by the head.

PRESSURE DYSPLASIA

This is a failure of development in the past combined with a deficient power of growth in the future A familiar example is the deficient growth on the inner side of the foot and ankle in a clubfoot deformity, due to the compression of these parts before birth, the result being the notorious tendency of such feet to turn into varus as development proceeds, even after marked valgus has been produced by treatment.

DISUSE DYSPLASIA

This effect is due to lack of the stimulus that only active movement of parts in their correct relationship can give. The best example is the shallow acetabulum and poorly developed head of the femur seen in congenital dislocations of the hip which have been reduced too late, and so have been deprived of the normal stimulus of use in the period of most active growth.

Now, if there is this failure of proper development to start with, and if development depends on active movement, it is surely inadvisable to try to correct such deformities by immobilization and consequent abolition of use. Apart from the lack of the stimulus of growth provided by action, immobilization of a bone-joint-muscle system has another result, whose exact mechanism is in doubt—the stiffening of joints by periarticular infiltration and fibrosis According to the reasoning I have used, this stiffening of joints, as well as atrophy of muscles, can occur before birth as well as afterward. It is obvious that it should be avoided if possible.

The present almost universal method of immobilization is by means of plaster of Paris molds or casts. They have the advantage of fitting the body exactly and being (perhaps deceptively) easy to apply. But apart from the general disadvantages of immobilization which have been set out, plaster of Paris has certain particular drawbacks, especially when used on the very young:

1. Being hard and rigid, it is very apt to produce a sore over a bony point.

2. Being inextensible, it may produce disasters if put on too tightly or if swelling occurs beneath it. This is because of a vicious circle in which tightness produces swelling

from interference with venous and lymphatic circulation. This interference produces more swelling, and so on, till complete stasis and gangrene of greater or less extent ensue.

3. Being absorbent of fluid, it soaks up urine, and as a result becomes both offensive and inefficient.

It needs to be stated that at the present time the range of generally accepted splints is remarkably inefficient. For instance, the commonest of all elements in deformities of the foot is equinus, yet there is no classic splint that will keep the opposite position of calcaneus; the stock tin shoe will not even keep the foot at a right angle to the leg. There is no classic splint that will correct in infancy that congenital postural scoliosis which later on turns into the intractable and crippling condition of "idiopathic scoliosis"; no device that will control a torticollis short of immobilization in a most uncomfortable plaster cuirass; no really efficient splint to correct bandy legs or knock knees; nor one to correct wrong pointing of the feet. The consequence is that vast numbers of unnecessary osteotomies and divisions of the tendo achillis are performed which could be avoided easily by better surgical mechanics.

Now, to design splints that will control movement without eliminating it completely, I think that the important principle is that as the muscles and the joints work in a series of lever actions, the best way to control them would be by means of a counterbalancing system of levers. This sounds platitudinous, yet it is not easy to cite a classic splint that works on this principle; and the modifications made to my splints

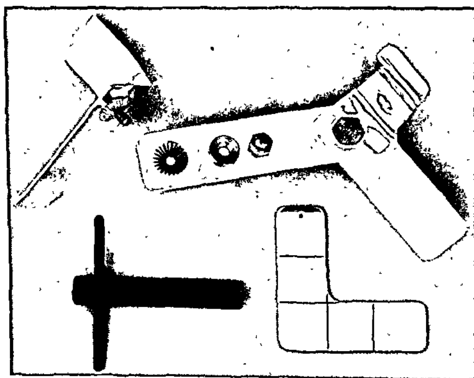


Fig 1. The hobble splint showing the left footpiece connected to the crossbar, and the serrated cone friction joint on the right side, with the washer and the nut which fix it in any desired position. The metal blank for the footpiece also is shown, marked in order to show its proper proportions of 5 equal squares arranged in an L-shape. To correct a clubfoot, the footpiece finally is rotated outward until the affected feet are at right angles to the sagittal plane (pointing due east and west). The "box-type" spanner shown is the most convenient form for adjusting the friction joints.

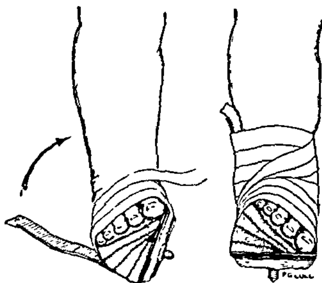


FIG. 2. Showing the correct use of the legpiece of the hobble splint to produce valgus of a clubfoot. Note that a thick pad of several layers of adhesive felt is stuck to the foot (not to the splint) just under the cuboid bone. If this is done properly, the legpiece should stand out from the leg as shown. Then when the legpiece is strapped in to the leg, the whole foot, including the heel, is swung into valgus. It is particularly in regard to the control of the heel that it is a bad mistake to remove or shorten this legpiece.

suggest that many surgeons do not see them as lever systems but as "cases" of the type of tin shoes, etc.

The next point to consider is how the forces produced by the system of levers in the various splints are to be applied. I have said already that the molding forces which are assumed to have caused the deformity should be imitated in correcting it. The characteristics of their pressure are pulsation, distribution and lack of friction. The pulsation can be supplied by the instinctive movements of the child, which are particularly powerful and persistent during infancy; an older child or an adult may lie quiet in a splint, but a baby responds to the force it exerts by fighting it continually, even if contentedly, all the time it is awake.

The softness and the distribution of the correcting pressure are obtained by using adhesive plaster and, in particular, adhesive felt as the means of applying it (see Fig. 7). In addition to adjusting themselves to any bony points, these preparations eliminate friction, since they do not move on the skin. In this way they avoid a cause of soreness which is only too well known to anyone who has done a long march.

THE HOBBLE SPLINT FOR CLUBFEET

This splint (Fig. 1) has 3 levering actions in the feet, 2 of which are eliminated partially or completely in the common modifications of it. The first is the connection between the feet which enables one of them to be controlled by the other in certain planes, particularly as regards rotating them inward or outward in relation to the body as a whole. The second is the control of equinus, gained by the side lever or legpiece so frequently cut off or shortened to an extent that stops its action. The third is the control of varus, gained by the same outside lever, assisted by packing adhesive felt under the outer side of the foot. This is perhaps the most important of all its actions, as it is the persistence or the recurrence of varus which is the main element in crippling from the deformity of the clubfoot. If applied properly to give pressure under the cuboid, the packed felt will prevent the disastrous deformity of rocker foot. (Fig. 2.)

The main modifications of this splint which I know of, and none of which I regard as an improvement, are:

1. The removal or the cutting down to a rudimentary size of the outside lever or legpiece. Apart from its function of forcing the ankle into valgus, this part of the splint gives the main grip on the limb to keep the heel down; therefore, it is rather annoying to find a surgeon who has abolished this part of the splint criticizing it on the grounds that it will not hold an infant's foot.

2. The bending of the connecting bar in

order to give valgus to the ankles. This is very much inferior to the proper use of the outside lever combined with adhesive felt.

3. The lengthening of the connecting bar. I cannot see the point of this, though I have known it to be done with the idea of giving abduction of the hips and so correcting their congenital dislocation. In my opinion, abduction of the hips in this condition needs to be combined with extreme flexion to bring the head down and forward into the acetabulum.

4. The application of the splint with the footpiece fixed to the connecting bar. It is hardly necessary to point out the complete failure to understand the mechanics of the various levers shown by this.

5. The padding of the splint instead of the child. This already has been condemned as not eliminating friction.

6. The adding of a spike behind the sole plate. There can be no point in this if adhesive plaster and not bandages is used, as the former certainly will not slip off the angle on the proper model. It seems to me the only effect will be upon the sheets on which the child lies.

7. The shaping of the solepiece to correspond to the foot. This very common modification shows that those who make it do not understand that the projecting exterior-anterior angle of the proper oblong shape is there for the purpose of pulling the forefoot toward it, and so correcting metatarsal varus.

8. The fitting of universal joints to the footpiece holding the normal foot in unilateral cases. This is a curious modification which does away with the fundamental principle of the splint, the control of one foot by means of the other. In the version I have inspected, the universal joint permitted the baby to twist its deformed foot completely in the footpiece, as it would have done had there been no connection to the other foot at all.

9. The complicated fixings of the footpiece to the connecting bar. The device of pinning a serrated cone on the footpiece into a corresponding depression in the bar is stronger, cheaper and more efficient than any of the many complications that I have seen.



FIG. 3. Case of severe clubfoot, aged 12 days.

10. The use of thumb-screws, instead of nuts tightened to fix the footpieces by a cross-handled spanner.

11. Wrong curves to the legpiece. When this part of the splint is the proper length, it is important to have it bent in the proper curve. This bulges outward to clear the external malleolus, which otherwise is liable to develop pressure sores, then comes inward to meet the outer side of the leg, and finally bends outward sharply at its top to avoid digging into the skin at this point.

12. Wrong proportions of the splint in general. The simple rule that the footpiece is composed of five squares will give its proper shape (see Fig. 1).

THE EQUINUS SPLINT

This removable splint has many functions, depending on its power of reversing the very common equinus element in foot deformities and combining this with turning the feet in or out and into valgus or varus.

In addition to being absolutely indispensable to combating the differential growth of the inner and the outer sides of the ankle



FIG. 4. The same case as Figure 3, aged 18 months, wearing the equinus night splint with jointed connecting bar. Note the 3 actions—to turn the feet outward, to pull them up into calcaneus and to twist them into valgus. This splint must be worn at night for at least 5 years to counteract the differential growth of the inner and the outer sides of the foot.

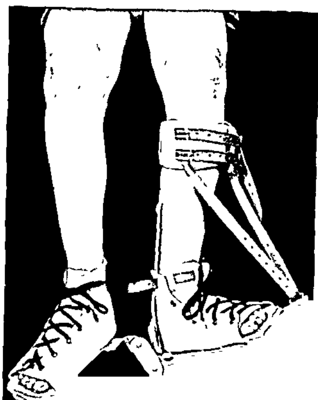


FIG. 5. Equinus night splint arranged for a left unilateral clubfoot, showing the use of a simple bootee on the normal foot to give a purchase for the turning out of the affected one. This model is a recent improvement on the one shown in Figure 4, the posterior strut being fixed to the soleplate of the bootee by means of a double hinge universal joint instead of to the ankle strap.

which turns the corrected clubfoot from valgus to varus, this splint is extremely useful in three other conditions:

1. The very common form of poliomyelitis that causes paralysis of the extensors and the peronei.
2. The invariable setting down into the equinus of the spastic foot.
3. The rare but annoying contraction of the calf muscles that some strong normal children develop owing to walking persistently on their toes. Vast numbers of divisions of the tendo achillis and rotation osteotomies can be avoided by the use of this splint. The splint consists of:

A. An open-ended bootee to hold the foot. The laces should not come so high on the ankle as to become tight when the foot is pulled up into equinus

B. A padded ankle band separate from the bootee which can be pulled tightly round the thin part of the ankle.

C. An extensible metal strut running up the back of the leg and holding up:

D. An upper band, which fits fairly loosely round the calf just below the knee and affords an anchorage for:

E. A strap which passes through and pulls upward:

F. A lever on the front part of the underside of the soleplate which can be rotated and fixed so as to pull the foot either straight upward, into valgus, or into varus as the condition demands.

G. A connecting link with two joints which permits the feet to be rotated either inward or outward in relation to the body as a whole, by means of joints similar to those in the hobble splint (Figs. 4 and 5).



Fig. 6. Another view of the latest model equinus splint, showing the fixing of the extensible posterior strut to the solepiece by 2 hinges at right angles to each other.

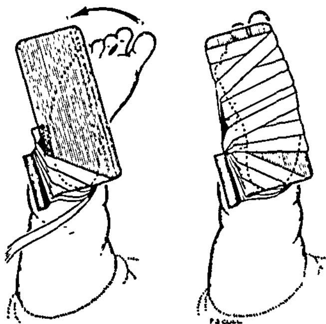


Fig. 7. Showing the correction of a metatarsal varus by means of the footpiece of the hobble splint, there being no need for the connecting bar in most of these cases. First, the foot is bandaged to the legpiece so that its anterior part projects inward from the solepiece, being held so by a thick pad placed over the apex of the curve of the deformity. Then the foot is bent into a reversed curve over this pad as a fulcrum and bandaged into position with adhesive tape.

Modifications

It is not yet well enough known to have many modifications made of it, the commonest being the addition of a strap over the instep to keep the heel down. This has the not inconsiderable disadvantage of stopping the foot from being pulled up into equinus, which is the main purpose of the splint. It comes from a failure to see that the heel is kept down by a tight grip on the leg just above the swelling of the malleoli. This, in turn, may come from a very common fault in manufacture, which is the placing of the ankle band so low, in an attempt to save leather, that it comes on the malleoli instead of well above them.

METATARSAL VARUS SPLINT

This is simply the footpiece of the hobble splint. Its application is shown in Figure 6. It is obvious that if the shape of the soleplate were modified to that of the normal foot, the whole action would be lost.

METATARSAL VARUS NIGHT SPLINT

This is a simple bootee with a curve of the sole which reverses the original curve of the foot and a strap which holds the foot to that curve (Figs. 7 and 8).

METATARSAL VALGUS SPLINT

This is the footpiece of the hobble splint again, but left in a flat L-shape instead of having the legpiece bent up at right angles (Fig. 9).

METATARSAL VALGUS NIGHT SPLINT

This is a bootee with an inner lever and strap to bring it into the leg and, in consequence, keep the ankle in full varus.

INFANTILE SCOLIOSIS SPLINT

According to the theory of the intrauterine production of certain deformities, the intractable and crippling condition of "idio-



FIG. 8. A pair of metatarsal varus feet that have been corrected in the way advised. Note the deep indentations, which can be made safely by pressure with adhesive felt. If similar pressure were put on by plaster of Paris, the result would be a deep slough.

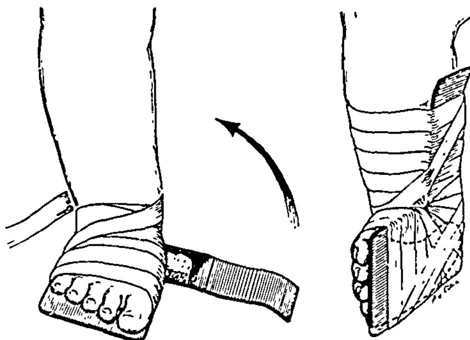


FIG. 9 Correction of a metatarsal valgus talipes in an infant. The splint used is the same L-shaped piece of *aluminum as in the hobble* splint, but without the bending up of the legpiece. First the foot is strapped to the solepiece with a small pad of adhesive felt under the prolapsed tarsal bones of the instep, then the ankle is forced into full varus by bringing the legpiece up to the leg.

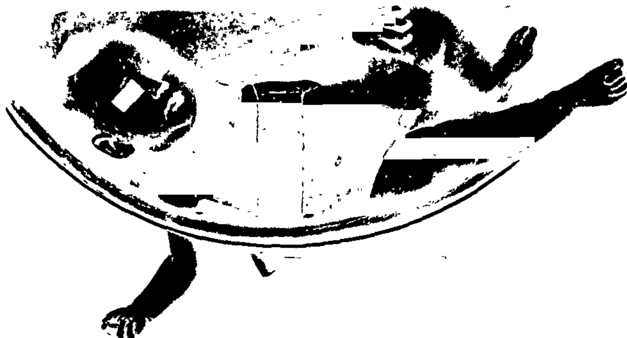


FIG. 10. Scoliosis splint, showing how original scoliosis curve, convex to the left, is kept reversed although the child can move freely.

pathic scoliosis" is a congenital postural scoliosis, due to the child's lying in utero with a slight lateral bend in the spine. Usually it is undiagnosed during its early, inconspicuous and curable stage, and the "plaster bed" that is the stock corrective is difficult to make, soon becomes small, and at best is a very inefficient mechanism to reverse the deformity.

The splint shown in Figure 10 consists of a flat plate of aluminum covered with sponge rubber. Along one side of it is a curved wall of similar construction, into the curve of which the baby is held by a broad padded band adjusted to the apex of the original scoliotic bend. There are two holes in the wall, toward either end of it, and the appropriate arm of the child is brought through one of these to lock it in position. In this apparatus the child can lie comfortably and perform almost any movement, with the exception of getting its spine out of a curve which is the reverse of the original one. Also, it can be carried about easily, which is a point of considerable importance.

MODIFICATIONS

The only one I have encountered so far is the straightening out of the curve in the wall where the child's head comes. This does away with a good deal of its effectiveness.

HIP SPLINT FOR CONGENITAL DISLOCATION OF THE HIPS

The idea on which this splint is designed is that the head of the femur has been pushed backward out of the acetabulum by pressure on the knee with the thigh fully flexed in the intra-uterine position. To get the head back into position, it is necessary first to flex it fully on the body and so bring the head back into the track along which it was dislocated. Then the actual primary reduction is performed by pressure upward and forward on the greater trochanter, while the child lies on its face, thus reversing the original thrust responsible for the displacement. This thrust along the axis of the femur gives much easier reduction with far less



FIG. 11. Posterior view of splint for congenital dislocation of the hip, showing the pad of sponge rubber over the sacrum, which acts as a fulcrum to enable the knees to be pulled backward, and the braces, which stop the hips from being extended.



FIG. 13. Case of congenital sternocleidomastoid torticollis.

pressure on the femoral epiphysis than the classic levering actions of Lorenz. Once the head is in the acetabulum it can be kept there, because the only way it can escape is backward, and this can be prevented by complete abduction of the knee, which pushes it forward on the fulcrum of the iliofemoral band. So long as the knee is kept right back, a fair amount of movement is permissible, which works the head into the socket and stimulates its growth.



FIG. 12. Anterior view of splint for congenital dislocation of the hips.

The splint consists of a bar, extensible to allow adjustment with growth, and padded malleable metal rings to fit the thighs at either end. Where the bar passes over the spine a large sponge rubber pad is fixed on it, and it is prevented from slipping down by simple braces of woven fabric over the shoulders (Figs. 11 and 12).

MODIFICATIONS

1. I myself have changed the original position of the bar from the front of the body to the back, to give greater abduction of the thighs.[2]

2. Superficially similar forms of splint are made with a rigid corset round the body, which does away with the powerful, though limited, movements of the thighs which are *the most important factor in developing the* hip joints.

3. There are versions with small wheels under the thigh bands so as to allow the child to skate about face downward. This does away with the powerful action of the

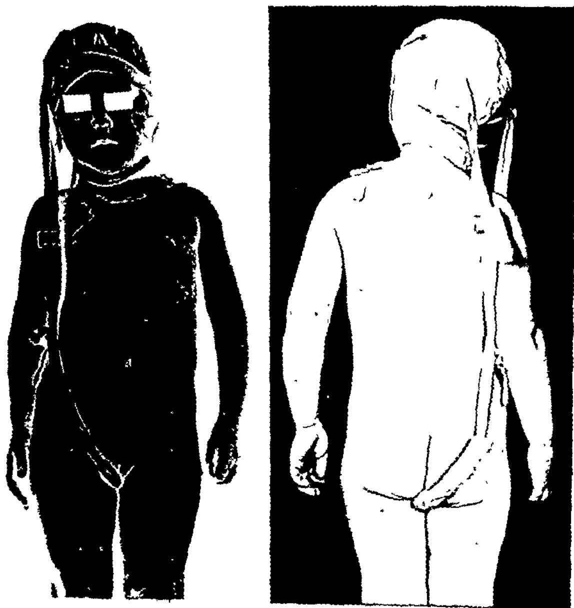


FIG. 14. (*Left*) This is the same case as in Figure 13 a week after open division of the upper end of the sternocleidomastoid combined with tenotomy of *the lower end.* Note the inextensible band of adhesive tape round the head which stops the linen cap to which the straps are fastened from slipping off. (*Right*) Back view of the *torticollis* harness, showing the padded webbing strap passing round the neck, crossing over the ear, and the 2 ends fixed to the linen cap in appropriate position by safety pins.

thigh muscles which is produced by efforts to crawl and to walk in the splint.

TORTICOLLIS CONTROL

I treat the form of torticollis caused by contraction of the sternocleidomastoid by dividing this muscle through an open incision at its upper end, and by a tenotomy just above the clavicle. In this way one gets very free relaxation, with inconspicuous scarring. Afterward it is necessary to hold the neck in the reversed position while permitting free movement, and so gaining the muscle balance which is so important to maintain correction. A linen cap is made to fit the skull closely, and this is fixed in position by binding it round by inextensible sticking plaster, in the line of the forehead and the occiput, below the greatest diameter of the skull so that it cannot be pulled off. Then a padded fabric strap is passed round the neck on the opposite side to the contracted muscle and brought over the top of the head, being fixed by safety pins to the linen cap in such a way that one end hangs down in front and one behind. Next, these ends are buckled to the ends of a similar padded strap passed through the crotch of the child, and crossed below the axilla. By tightening them up (Figs. 13 and 14) it is possible to correct both the scoliosis and the rotation of the cervical spine, and a fortnight in this device, moving about freely, establishes permanently a normal carriage of the head. It is important, both psychologically and physiologically, that the child wake up from the anesthetic with the position of the head corrected in this way; to adjust the position of the neck with a new wound in a conscious child is neither kind nor effective.

REFERENCES

1. Browne, Denis: Congenital deformities of mechanical origin, Arch. Dis. Childhood 30:149, 1955.
2. ———: Congenital dislocation of the hip, Proc. Roy. Soc. Med., Section of Orthopaedics 41:388-390, 1948.

Apparatos Orthopedic a Movimento Controlate

Summario in Interlingua

Le presente articulo stipula que certe deformitates congenite es causate per fortias mechanic que age super le feto in utero e que iste hypothese de causation permitte le derivation de importante principios therapeutic. Iste principios es (1) que le fortias usate pro reducer un certe deformitate deberea esser simile al fortias que supponitemente ha causate lo e (2) que il es indispensabile de permitter active movimentos durante le processo de correction a fin de contrabalanciar le effectos inimic del prenatal immobilisation e displaciamento del membros in question.

Pro applicar iste principios, varie apparatos orthopedic de invention original es describite, insimul con le currente variationes e modificationes de illos. Iste modificationes es criticate, e rationes es presentate pro que illos non pote esser considerate como meliorationes.

Le conditiones describite es

1. le tres varietates standard de modulation del pede, i.e.
   a talipede,
   b. metatarso var,
   c. metatarso valge,
2. dislocation congenite del coxa,
3. congenite scoliosis postural (del qual il es asserite que illo es le prime phase de "scoliosis idiopathic"), e
4. torticollis sterno-mastoide.

10

Congenital Dislocation of the Hip — Its Causes and Effects

WILLIAM K. MASSIE, M.D.*

Two hundred years after Robert Boyle postulated the hidden importance of fermentation, Pasteur proved it. Four years after Marie Curie postulated the existence of a new element, radium, she and her husband proved it. Postulation based on facts has been the parent of most significant advances in any field; hence, it is the apologia of every student of this most interesting and perplexing orthopaedic condition in offering or supporting an etiologic explanation. If such an explanation should be compatible with all the known facts, it not only simplifies the task of teaching but also provides a scaffolding from which rational treatment can be constructed.

If the many etiologic suppositions appearing in the literature in the past century were to be discussed here, the purpose of this chapter would be defeated. Rather, evidence will be given to support the single simple supposition that a pathologic relaxation of the joint capsule exists, and that all other pathologic changes develop subsequently as the result of simple mechanical stresses. Jones and Lovett,[19] in making this supposition, differentiated this condition from the congenital dislocation accompanying multiple anomalies that were thought by them to be primary. Howorth,[16] more recently, has presented this supposition as the explanation compatible with all the findings.

Whether capsular relaxation is truly genetic or merely acquired *in utero* and early infancy as the result of hormonal, dietetic, metabolic or mechanical factors will not be discussed, as no convincing data on which to base an opinion is known to the author.

*Lexington, Ky.

The embryologic studies of Strayer[35] and later Gardner[11] have demonstrated that the hip joint first appears as a blastemal condensation in the limb bud about 5½ weeks after conception, together with the associated muscles and nerves in definitive positions. No dislocation is possible at this stage, as all components of the hip joint develop from a single cartilaginous column. At 7 weeks the embryonic period ends, with all important structures assuming their adult relationship. Therefore, any positional rearrangement occurs in fetal life or afterward. Normally, in the embryonic period, a joint will develop independent of any mechanical factor, even when separated completely from the embryo. Hence, mechanical forces become effective only after the embryonic stage.

CAPSULAR RELAXATION AS RELATED TO THE COMMON FINDINGS IN CONGENITAL DISLOCATION OF THE HIP

Geographic incidence of the condition is striking. In the southern European countries, where it is common, frequently it is associated with the calcaneovalgus feet, but in the Negro so commonly affected by liga-

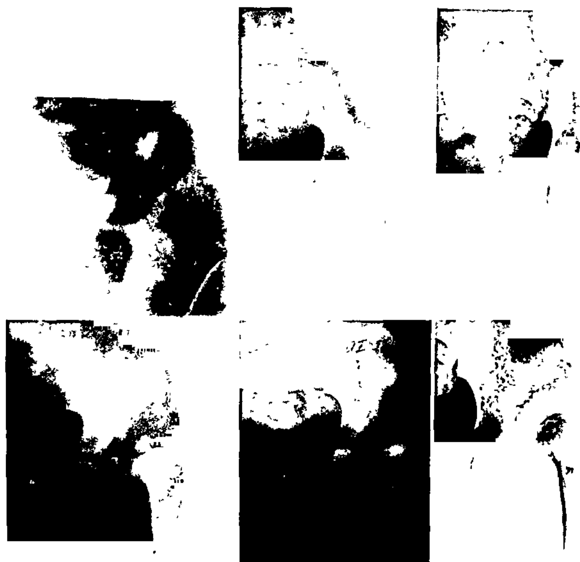


FIG. 1. Types of acetabular roof obliquity. *(Top 3 illustrations)* Hips at 2, 4 and 5 years. There is a false acetabulum in each case, and the acetabular roof obliquity is comparable with that of a newborn infant. The dislocation pre-existed weight-bearing and occurred rapidly, presumably due to a very relaxed capsule. The acetabular roof failed to develop after the head left the acetabulum.

(Bottom 3 illustrations) Hips, at 2, 3 and 7 years. The "notching" or bilabiation of the acetabular roof is present, and there is no well-developed false acetabulum. The dislocation occurred gradually, assisted by weight-bearing due to an only partially relaxed capsule. At *bottom, right* the anterior and the posterior lips of the acetabular roof are developed clearly, and there can be seen between them the furrow formed by the dislocating head.

mentous relaxation of the feet it is comparatively uncommon.

Sex Incidence. While dislocation occurs approximately 6 times as often in girls, dysplasia of the joint is only slightly more common in girls (Faber).[8] On a purely mechanical basis, this should be expected, as the plane of the acetabulum is more nearly vertical and the fossa more shallow in girls. Combined with the conventional fetal position, any degree of capsular relaxation could initiate the train of mechanical events discussed below.

Bilateral Incidence. This statistic varies with the observer and the locality from which he reports. Scaglietti[33] encounters

bilateral involvement in 54 per cent of his series. This does not take into account those patients with unilateral dislocation and contralateral dysplasia that occurred in 20 per cent of 35 cases reported by the author.[21] Still, there is an appreciable number with a normal-appearing hip on the contralateral side. This apparently is inconsistent with the capsular relaxation supposition. Alterations in the positions of the two extremities *in utero* may explain this variation adequately.

DISLOCATION IN UTERO

Though rare, this condition has been noted on routine prenatal roentgenograms. Ortolani[28] reported 2 fetuses aged 6 months and 7 months examined at autopsy. In both cases a ligamentum teres was found present bilaterally. In one fetus the bony acetabular roof appeared less oblique in the roentgenogram on the dislocated side than on the contralateral side. This showed evidence of capsular relaxation and subluxation. *Dislocation before the weight-bearing age rarely is* reported, though dysplasia or subluxation is recognized with progressive frequency due to the emphasis on early signs and symptoms. If dislocations existed commonly before the weight-bearing age, they too would be seen more frequently.

ROENTGENOGRAPHIC ANATOMIC VARIATIONS EXPLAINED PRIMARILY BY CAPSULAR RELAXATION

Acetabular roof obliquity, as seen in the roentgenogram, does not indicate the actual support provided by the acetabulum, since below the age of 3 years the cartilaginous portion is considerable. However, the bony acetabular roof is a good index of resistance to upward displacement, since it ceases promptly to decrease in obliquity when the normal femoral head pressure is decreased (Fig. 1, *top 3 illustrations*). If capsular relaxation permits rapid and complete dislocation of the head, the bony acetabular obliquity remains at the angle that it had attained at the time of dislocation, and a false acetabulum is formed above (Fig. 1, *top 3 illustrations*). If capsular relaxation increases gradually as the result of increased mechanical stress, there will be a gradual displacement of the head upward with the production of a furrow through the acetabular roof known as "notching" or "bilabiation" (Fig. 1, *bottom 3 illustrations*). This gradual displacement seems to be accompanied by more severe molding of the femoral head than is noted in the more abrupt dislocations. The author has not seen this latter condition in a child who was not weight-bearing when the diagnosis was made. The acetabular appearance and the head deformity in the abrupt type are simulated closely by poliomyelitis complicated by dislocation on attempted weight-bearing (Fig. 2, *left*), whereas the more gradual type is seen routinely in the dislocations complicating severe spastic paralysis (Fig. 3, *left & right*).

If the cessation of development of the acetabulum in the abruptly dislocating type or the notching of the acetabulum in the gradually dislocating type is a genetic fault and not the direct result of malapplied mechanical stress (Wolff's law), how can the consistent response of the acetabulum in the early subluxated hip to adequately maintained reduction be explained? Putti[29-31] demonstrated this consistent response when diagnosis and treatment were begun before the first year. Subsequently, others have corroborated his results. It is irrational to suppose that the replacement of a femoral head in anatomic position could reverse a genetic fault that caused the original subluxation or dislocation. The elongation of the ligamentum teres is another anatomic finding resulting directly from capsular relaxation. Occasionally this structure is absent, presumably due to attritional effects of



FIG. 2. *(Left)* Poliomyelitis in infancy; present age 25 years. Residual adductor contracture permitted dislocation on weight-bearing with stretching of the capsule. Compare with *(right)* congenital dislocation at 10 years of age. Note the acetabular roof furrow and bilabiation, the deforming head and the severe anteversion (patella forward).

weight-bearing, but more frequently it is attenuated and presents a formidable obstacle to replacement of the femoral head in an acetabulum already partially filled with dense connective tissue.

ANATOMIC FINDINGS SECONDARY TO THE DISLOCATION PER SE

Once the normal intra-articular pressure is decreased by gradual outward displacement of the femoral head, the decreased pressure results first in *actual thickening* and subsequent ossification of the triradiate cartilage and then in proliferation of a dense fibrous tissue floor.

Subluxation also is accompanied by *retarded epiphyseal ossification*. The center of ossification appears later and remains constantly smaller than the normal epiphysis. If its appearance is made and subsequently disappears or is not made at all by the 18th month, one must consider the possibility of vascular damage to the epiphysis subsequent to some abnormal trauma The ossification of the triradiate cartilage and the ischiopubic synchrondrosis also is retarded.

Dunlap, Shands *et al.*[7] found the normal femoral torsion in 16 children between the ages of 3 and 8 months to average 31°. In 29 patients and 40 dislocated hips the average torsion was 51°. The age of the patients was not listed. In 3 dislocated hips, measured before the patients began independent walking, using the method described by Crane and Ryder,[32] the author found no femoral torsion exceeding 40°. Since this



FIG. 3. *(Left)* Spastic quadriplegia at 4 years of age. The patient stood with legs adducted and with severe internal rotation. Hip is in the process of gradual dislocation. *(Right)* Same patient as at *left* at 8 years of age. The dislocation now is complete; the acetabular furrow has formed with the typical bilabiation. Compare with Figure 2.

amount of torsion is approached by a normal femur in the first 6 months, it is deduced that in the presence of a dislocation the normal process of derotation does not take place.

ANATOMIC FINDINGS SECONDARY TO WEIGHT-BEARING ON A DISLOCATED HIP

Progressive femoral torsion (anteversion) appears to result from, and not to be a cause of, congenital dislocation when weight is borne on the dislocated hip. The following groups exceeded 50° internal femoral torsion (anteversion) when estimated at open reduction (Fig. 4):

59 per cent of 17 hips from 2 to 3 years
100 per cent of 9 hips from 3 to 4 years
82 per cent of 11 hips from 4 to 5 years
100 per cent of 13 hips from 5 to 8 years

Four hips in patients over 12 years measured over 70° by the above-mentioned roentgenographic technic (Fig. 5), and a 40-year-old adult with bilateral uncorrected dislocated hips measured 80° by the same method (Fig. 6). Femoral torsion is an internal twist of the shaft of the femur (Hibbs),[15] which, it is believed, results from the constantly repeated contraction of the thigh adductors and medial hamstrings exerted on the externally rotated lower extremity fixed at the hip in this position by the contact of the femoral head with the wing of the ilium and by the total loss of internal rotators resulting from upward displacement of the hip and relaxation of the gluteus medius. A similar femoral torsion of equal





FIG. 4. Demonstration of progressive anteversion or femoral torsion with weight-bearing. 2½ years old—ambulatory 1½ years. The right (dislocated) hip shows an anteversion or femoral torsion of 50° compared with 30° on the left. (Technic: Crane & Ryder) The angle formed by the shaft and the neck in this view indicates the degree of torsion.



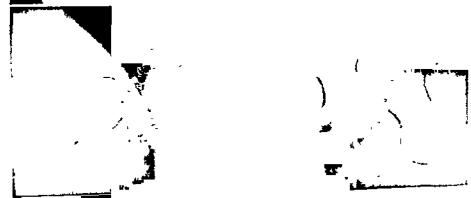


FIG. 5. Demonstration of progressive anteversion or femoral torsion with weight-bearing. A 12-year-old girl ambulatory since childhood, with no attempt at correction of the dislocation. A femoral torsion of 70° is present.



FIG. 6. Demonstration of progressive anteversion or femoral torsion with weight-bearing. The mother of patient in Figure 5, aged 44. A femoral torsion of nearly 90° is present.

severity can be demonstrated in a severe spastic paraplegia with adductor spasm and loss of functioning gluteus medius, but in these patients the torsion is aided by a peculiar sitting position habit.

Failure of the normal acetabular roof development is invariable when there is no femoral head in the socket to exert a normal head-acetabular pressure. When left untreated over many years, a new incomplete socket is established along the side of the ilium from pressure, but the original acetabular fossa disappears almost completely. If the dislocation has been of the gradual type, a deep notch in the acetabular roof



FIG. 7. Development of coxa valga. A 22-year-old female with identical bilateral involvement. Capsular relaxation permitted only moderate subluxation. The acetabular roof obliquity measures 20°, which is the average obliquity of a 1-year-old child. The patient developed a severe coxa valga, which in this illustration is accentuated by femoral torsion (taken with patella forward). Note that the epiphyseal line is perpendicular to the long axis of the femur. This hip was far more symptomatic than those shown in Figure 6.

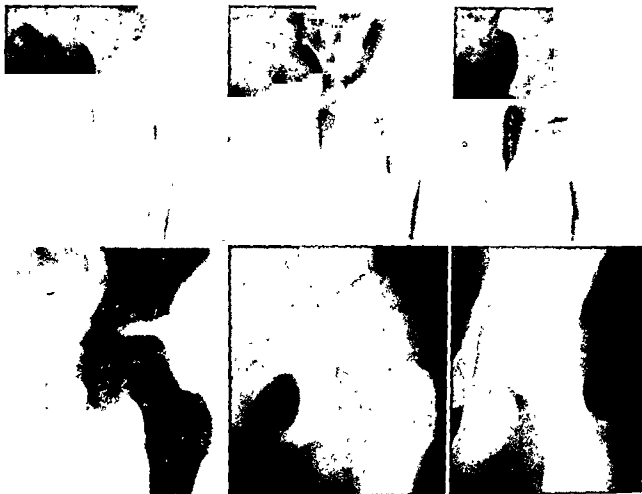


FIG. 8. Adequate acetabular response to reduction. *(Top, left)* Aged 18 months; note severe sloping of acetabular roof and anteversion present. *(Top, center)* Aged 2 years. Adequate reduction maintained by abduction 6 months; anteversion corrected by osteotomy. *(Top, right)* Aged 4½ years; acetabular response is definite but slow. *(Bottom, left)* Aged 6½ years. *(Bottom, center)* Aged 7½ years; reduction is maintained, but the acetabular response has been slow. *(Bottom, right)* Aged 23 years; adequately formed hip. There is excellent function.

can be identified, as mentioned previously, by the remains of the anterior and the posterior rims of the acetabular roof that persist (Fig. 2, *right*).

The femoral head, unstimulated by any pressure from above, fails to develop and remains small and somewhat irregular. However, it does not develop the gross irregularities commonly noted in femoral heads affected by avascular changes (Fig. 2, *right;* compare with Fig. 14)

Coxa valga is not present when dislocation develops rather rapidly in the first year of weight-bearing. This is in marked contrast with the congenital subluxation that results in a bony resistance to weight-bearing by an insufficient acetabulum and a gradual outward displacement of the center of rotation of the femoral head. In such femurs the medial trabeculations almost parallel the long axis of the femur, and the patient walks with a gluteus medius limp more pronounced than is noted in the completely dislocated hip (Fig. 7). Such hips usually are much more painful than those completely dislocated. As shown by Inman,[19] the development of coxa valga results from a shift of the weight-bearing or resultant force acting through the neck of the femur from an angle of approximately 155° with the

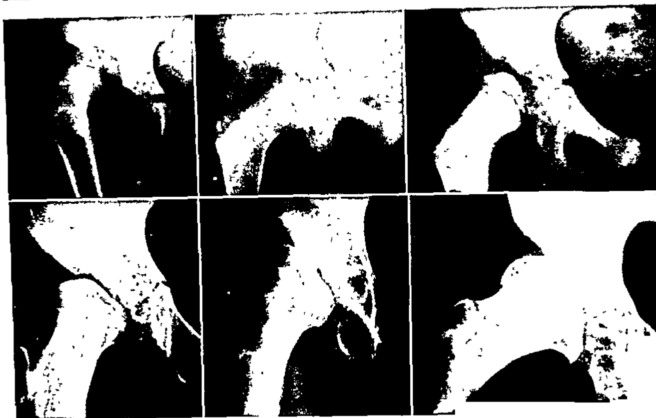


FIG. 9. Acetabular response to resubluxation. *(Top, left)* Aged 7 years. Notched acetabular roof; anteversion 80°. *(Top, center)* Aged 7½ years; open reduction with femoral osteotomy to correct anteversion has been done, but a shelf was not provided. Resubluxation is mild. *(Top, right)* Aged 8 years; resubluxation is pronounced. The acetabulum has responded only slightly with a decreased obliquity. *(Bottom, left)* Aged 11 years; subluxation has persisted, and the roof obliquity has remained unchanged. Note the position of the epiphyseal line perpendicular to the long axis of the femur. Coxa valga is developing. *(Bottom, center and right)* Aged 28 years; roof obliquity has remained unchanged since puberty. The hip is severely symptomatic. (Massie *et al.*: J. Bone & Joint Surg. 33A:179)

femoral shaft to more nearly vertical. This is accompanied by the marked abductor lurch, which is an attempt to bring the center of gravity directly over the femoral shaft. Since the epiphyseal plate remains perpendicular to the weight-bearing axis, it remains almost horizontal to the femoral shaft, and the neck grows in this axis (Fig 7).

Increased lumbar lordosis is a classic finding when bilateral dislocation is accompanied by a backward displacement of the femoral heads and a forward tilting of the pelvis.

ACETABULAR RESPONSE TO ADEQUATE REDUCTION

Putti[29–31] demonstrated that the simple abduction treatment was effective in producing a normal-appearing hip when instituted before the first year. The author, in a report with Howorth[26] covering a study of 58 hips all treated by open operation after the age of 1 year and followed to adult life, found no exception to this. *This study demonstrated that adequate reduction obtained and maintained would result in a normal acetabular response* up to the age of 8 years. However, over 3 years acetabular shelf (Fig. 8) procedures to accentuate this response seemed to be indicated progressively. We accept this premise as fact and offer it as the most potent rebuttal to the theory of genetic acetabular inhibition or "dysplasia" in such patients.

Putti[29–31] stated that bilateral hips after the age of 4 years and unilateral hips after the age of 7 years should be left unreduced.



FIG. 10. Acetabular response at 7 years aided by adequate reduction and primary shelf procedure. *(Top, left)* Aged 7 years; appearance almost identical with that in Figure 9, *top, left*. *(Top, right)* Open reduction, shelf procedure and rotation osteotomy provide an adequate reduction and acetabular roof support. *(Bottom, left)* Aged 14 years; acetabular support now is complete. There is no resubluxation. Presence of coxa vara is unexplained. *(Bottom, right)* Aged 26 years; there is an almost normal range of motion, and the patient remains asymptomatic. Compare this result with Figure 9, in which resubluxation was permitted to occur. (Massie *et al.:* J. Bone & Joint Surg. 33A:185)

Though the percentage of excellent roentgenographic results in older children in the above quoted series was only 17 per cent symptomatically in adult life 65 per cent were graded as excellent. Colonna[4] has advocated a capsular arthroplasty for this age group. One seems justified, then, in taking exception to Putti's dictum but recognizing in his statement what he implied. Hips over 3 years of age respond much more slowly to the regenerative stimulus of adequate reduction. The surgeon has a more difficult task not only in *obtaining adequate reduction but also in maintaining it*. If the latter is imperfect, it would have been better had the job been left undone (Fig. 9). In maintaining such a reduction, it is assumed that each condition that favors subluxation will be treated adequately: (1) open reduction if closed reduction cannot seat the head normally; (2) shelf procedure if the acetabular roof is insufficient to support the head; (3) rotational osteotomy if femoral torsion favors an unstable reduction; (4) prolonged immobilization until there is roentgenographic evidence of *normal restoration of the head-acetabular relationship*. This must be accomplished without the onset of avascular necrosis (Fig. 10).

FEMORAL HEAD CONTOUR RESPONSE TO ADEQUATE REDUCTION

The femoral head at open reduction often appears to be deformed. No measures need be taken to improve the symmetry, since even heads over 5 years of age tend to become spherical (Fig. 10). However, any degree of so-called aplasia or epiphysitis that indicates uniformly a vascular disturbance may



FIG. 11. Hilgenreiner's measurements. "h" is the distance between the "y" line and the highest metaphyseal point. "d" is the distance between the intersection of the "y" line with "h" and with the acetabular line. "a" is the angle of the acetabular roof. Note the difference in the measurement on the 2 sides: "a" and "d" are greater on the right, and "h" is smaller; this hip is subluxated.

progress to a grossly deformed head that deforms the acetabulum secondarily (Fig. 14).

Adequate reduction does not seem to alter an existing femoral torsion (anteversion) or coxa valga. It may be that since both conditions become severe only in the older patients, sufficient time before bone maturity is not available for the reversal of these findings.

DIAGNOSIS OF SUBLUXATION ROENTGENOGRAPHICALLY

Adequate reduction implies the absence of *any* subluxation. The diagnosis of gross subluxation at any age is quite apparent, but it has been stressed that even minor degrees of subluxation that persist after reduction of a dislocated hip may progress steadily to either redislocation or an unstable hip giving dire adult symptoms. *Any* degree of subluxation should be recognized and watched carefully by serial roentgenograms. Evidence of progression or persistence of subluxation should be met with immediate appropriate treatment.

Shenton's line (the curved line formed by the inferior margin of the femoral neck continuing along the interior margin of the superior ramus of the pubis) is useful in the recognition of dislocation, but it is an unreliable sign in mild and moderate subluxation It may appear broken, signifying subluxation when such is not present—i.e., severe flattening of the femoral head without outward displacement or a combination of coxa valga and anteversion—and it appears quite normal unless there is gross upward displacement of the head (Fig. 11). Hilgenreiner's measurements are more reliable, particularly in infants; but the normal measurements must be available if the dislocation is bilateral, and, if it is unilateral, care must be taken to ensure an accurate anteroposterior view of the hips and the pelvis. These measurements are not applicable after the capital epiphysis has developed completely. The author prefers to use a measurement first described by Wiberg[35] and later used by the author in conjunction with Howorth.[25] The Center-Edge angle of Wilberg is the angle formed with the horizontal by a line passing through the *center* of rotation of the femoral head and the *edge* of the acetabulum (Fig. 12). If a true anteroposterior view of the pelvis is obtained, the position of the femoral head and neck does not alter the measurement. It is ascertained quickly by the use of an overlay that determines accurately the center of the head when the epiphysis is well developed, and its relation to the edge of the acetabulum is read off in degrees. Use of this method in children below the age of 3 years becomes increasingly difficult. The author has found

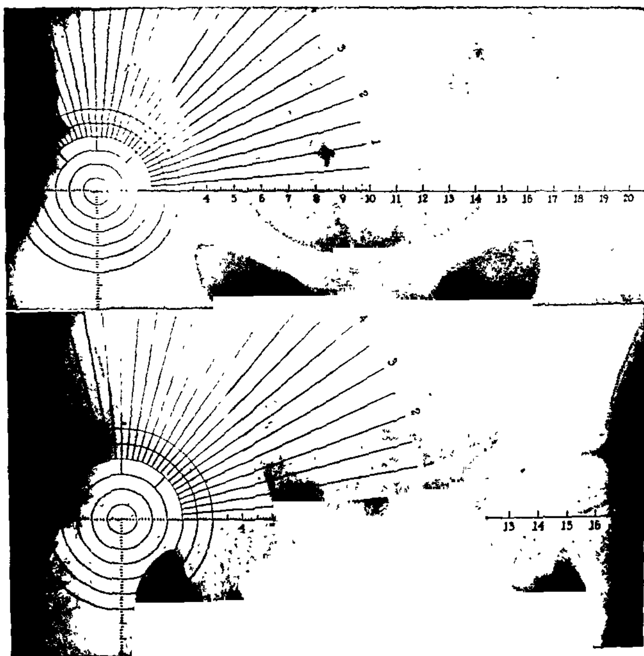


FIG. 12. Use of the CE angle in determining subluxation. *(Top)* Aged 12 years; the CE angle is 30°. The center of rotation is determined by superimposing one of the circular lines over the periphery of the head. The horizontal line must be placed symmetrically for the 2 sides of the pelvis, and the roentgenogram must be a true anteroposterior view. *(Bottom)* aged 12 years; the CE angle is 0°. Note that this angle can be measured with the hip in any degree of rotation or abduction adduction, as the center of the head is the one point that does not change with various positions of the hip. (*Bottom*—Massie *et al.*: J. Bone & Joint Surg. 32A:526)

that the center of rotation of the cartilaginous head lies at a point equidistant from the 2 poles of the metaphyseal line and just proximal to the metaphyseal border. However, one may calibrate easily the accuracy of his technic, since on a given patient the reading should be the same with the hip held in abduction, adduction, internal rotation or external rotation. Any lateral shift of the head usually is quickly apparent without reference to tables, but normal values for all ages have been published previously.[25]



FIG. 13. Vascular changes occur bilaterally in a girl treated by abduction for dislocation of the left hip only. *(Top, left)* Aged 1 year; dislocation on the left. *(Top, center)* Aged 14 months; closed reduction is adequate, attained by simple abduction in plaster. *(Top, right)* Aged 2 years and 7 months; 17 months after treatment began both epiphyses show evidence of vascular changes. *(Bottom, left)* Aged 4 years; changes more obvious. *(Bottom, center and right)* Aged 4½ years. Mild subluxation bilaterally; fragmentation of the epiphyses persists; patient being kept off weight-bearing until epiphyses appear to be normal.

VASCULAR EPIPHYSEAL CHANGES

These changes are easily discernible and of sufficient frequency to be familiar to all who treat this condition.[1,23] They have been described in detail elsewhere.[24] None of these changes has been observed in congenitally dislocated hips not subjected to some type of treatment, but they were noted in the contralateral normal hip of one patient subjected to closed reduction of the dislocated hip and bilateral cast fixation (Fig. 13). They were noted in 30 per cent of 58 hips reduced openly and in 45 per cent of 31 hips reduced by the closed method. They appeared first from 3 to 12 months after reduction.

The vascular changes are divided into 2 groups: those with epiphyseal fragmentation similar to that seen in coxa plana and those without fragmentation.

The first group (Figs. 14 & 15) are divided easily into 3 grades of severity: mild, moderate and severe. When followed to adult life, only those hips with mild involvement functioned adequately. Among the findings characterizing this group are: (1) initial epiphyseal fragmentation; (2) gradual flattening or occasional disappearance of the epiphysis; (3) convexity of the medial metaphyseal edge of the epiphyseal line; (4) gradual broadening of the metaphysis.

The second group, comprising 40 per cent of the series studied, differs from the first group in the first 2 findings: no fragmentation was noted, and only the medial pole of the epiphysis became flattened. The metaphyseal changes were the same. The adult results of the second group, though not good, were much better on the average than those of Group 1 (Fig. 16).

In this series 87 per cent of the male hips dislocated congenitally and 25 per cent of the female hips showed vascular changes. This sex incidence is comparable with that of coxa plana in an older age group. The constant relationship to reduction would seem

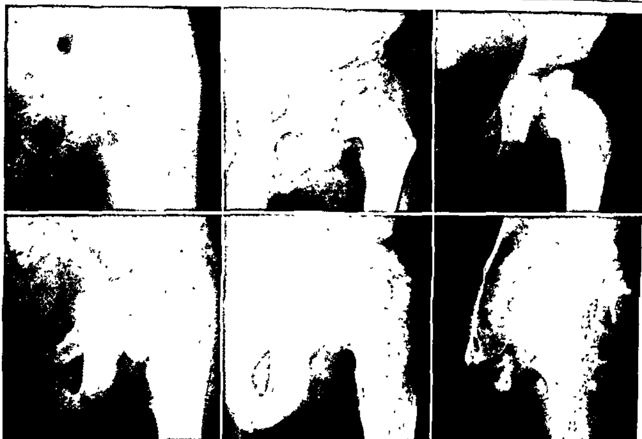


FIG. 14. A deceptive vascular epiphyseal change. *(Top, left)* Aged 14 months; acetabular obliquity and anteversion noted. *(Top, center)* Aged 22 months; slight fragmentation of the medial epiphysis, but the convexity of the metaphyseal edge already is apparent. *(Top, right)* Aged 3 years; reduction remains good, and the acetabular response is good. The vascular change no longer is apparent. *(Bottom, left)* Aged 6 years; progressive deformity of the head noted. *(Bottom, center)* Aged 11 years. *(Bottom, right)* Aged 23 years; marked head irregularity and cystic changes seem to be out of proportion to the minor vascular changes noted at *top, center.*

to implicate trauma in its production. Yet, with emphasis on gentle manipulation, these changes continued to appear in about the same percentage noted above. Preliminary traction was not employed in any of these cases, and Crego and Schwartzman[7] reported no such changes when traction was applied routinely. However, other observers using traction routinely[1] have continued to observe these changes. Adequate traction preliminary to any type of reduction deserves a thorough trial under the observation of many clinicians.

Excellent observations have been made on the vascular supply to the femoral head and neck.[2,20,36,37] However, in 1943, Wolcott[39] refuted the importance placed previously on the role of the ligamentum teres in the infant and the child but emphasized its role in the adult. Tucker[37] and, more recently, Trueta and Harrison[36] and Judet and Judet[20] have, by progressively more efficient injection methods, supported these conclusions. *Marked individual* variations have been noted uniformly, but it is agreed that while primarily cartilage, the epiphyseal ossification center receives its supply from the superior and the inferior capsular vessels, and that anastomosis with the ligamentum teres artery is not effected until this center enlarges to replace the intervening cartilage. In 20 per cent of the hips, no such anastomosis ever is noted. It is probable that recently employed radioisotope studies will give a far more detailed account of the vascularity of this region, its variability as to age and sex and its susceptibility to injury, both minor and severe. However, until we have such data at our disposal, we must be



FIG 16. Group 2 vascular changes (no epiphyseal fragmentation). *(Top, left)* Aged 3 years. *(Top, center)* Aged 3 years and 9 months; 9 months after reduction the medial lip of the epiphysis is flattened, and the metaphysis is rounded. *(Top, right)* Aged 5 years. *(Bottom, left)* Aged 6½ years; no subluxation, but flattening of the medial epiphysis persists. *(Bottom, center)* Aged 11 years; deformity of the head persists, but still there is no subluxation. *(Bottom, right)* Aged 22 years; mild subluxation and obvious head deformity. (Massie *et al.*: J. Bone & Joint Surg. 33A:290)

would serve only to confuse and reduce individual initiative, which is our greatest assurance of improved technics and results. An attempt has been made above to explain the various sequalae commonly encountered on a purely mechanical basis of cause and effect. If each failure can be explained, it will not be repeated. It is believed that no genetic factor exists that will compromise the results, provided that 3 simple requirements can be attained: (1) adequate reduction (anatomic restoration of the head-acetabular relationship), (2) maintenance of reduction, and (3) avoidance of vascular changes.

There is now sufficient evidence to state categorically that if the diagnosis is made within the first year of life and treatment is instituted promptly, a near-normal hip should result consistently. During this period, the maintenance of simple abduction seems to be sufficient, though some observers prefer to add internal rotation.[5,17,22] Mobile abduction splints are quite effective.[3,21] After this optimal period has passed, we approach rapidly an age in which preliminary adequate (skeletal) traction seems to be important to relax the surrounding tissues, so that after reduction is obtained (usually without any need for anesthesia), undue pressure is not exerted on the head. Reduction may be maintained by the method most familiar to the operator, either of a mobile or an immobile type, but of paramount importance is the roentgenogram, repeated frequently, indicating anatomic reduction.

The 45°-angle view of the hips and the pelvis, as described by Martz and Taylor,[21] modified slightly by using an 0.3 focal plane tube, seems to be a simple, effective technic. After the second year, depending on the roentgenographic and the clinical findings, one must choose between a closed or an open approach.[9,10,12,15,17,22] Usually this choice is made by either the success or the failure of the closed method to obtain and maintain reduction. After the third year, the acetabular roof is so deformed, femoral torsion is so pronounced and capsular and muscular contractures are so severe that the author elects the operative approach initially, preceded by skeletal traction and correction in stages of the secondary deformities incident to prolonged dislocation by open reduction with capsular plication, acetabular roof reconstruction and femoral osteotomy for torsion. The treatment after this age is highly individualized, but, as the age increases, the preparatory period prior to the initial reduction increases, interspersed with tenotomies and myotomies to reduce flexion and adduction contractures preliminary to traction.

Colonna's[4] capsuloplasty is an added procedure to be considered in the treatment of older age groups, particularly if the dislocation is unilateral.

The treatment of bilaterally dislocated hips beyond the age of 7 years is so fraught with difficulties as not to appear feasible to the author. Improved technics may alter this opinion later. However, unilateral dislocation is so rapidly symptomatic that one is justified in the attempt to restore head-acetabular relationships up to the age of puberty. Arthroplasty or fusion is the alternative.

SUMMARY

Congenital dislocation of the hip results from mechanical stresses applied to a joint in which a pathologic relaxation of the joint capsule exists, as proposed by Howorth[16].

All other pathologic changes, that is, slanting acetabular roof, femoral head deformities, femoral torsion and coxa valga, are secondary responses effected by an altered mechanical stress. The vascular changes commonly encountered in the epiphysis after treatment result from an individual relatively insufficient vascular arrangement of a specific hip that increases its susceptibility to even mild trauma. The changes are comparable with those encountered in coxa plana at an older age, and, if left untreated, produce a grave adult deformity. With the exception of these vascular changes, all deformities are reversible and amenable to treatment, though the difficulties of treatment increase rapidly after the second year and are so formidable after the age of 7 years that the treatment of a bilateral condition then seems to be inadvisable. Treatment in the early ages consists of the maintenance of abduction with or without internal rotation by the means most familiar to the operator until the normal head-acetabular relationship is stable. After this age the treatment becomes highly individualized, but there seems to be sufficient evidence to advocate skeletal traction as a preparatory measure to either the open or the closed reduction technics. Treatment at this age and subsequently should be undertaken only by the surgeon with considerable prerequisite experience or under his direct tutelage, since the entire future of the hip is in the balance.

CONCLUSION

If avascular changes can be obviated by extreme caution, every congenitally dislocated hip seen before the second year can be expected to terminate as a near-normal one, both anatomically and functionally, and each hip after this age can be expected to respond to anatomic reduction with a good permanent result if this reduction can be maintained at all times.

REFERENCES

1. Bost, F. C., Hagey, Helen, Shottstaedt, E., and Larsen, Loren: Results of treatment of congenital dislocation of the hip in

infancy, J. Bone & Joint Surg. 30A:454-468, 1948.

2. Chandler, S. B., and Kreuscher, P. H.: Blood supply of ligamentum teres, J. Bone & Joint Surg. 14:834-846, 1932.
3. Chuinard, George E.: Early weight-bearing and the correction of anteversion in the treatment of congenital dislocation of the hip, J. Bone & Joint Surg. 37A:229-245, 1955.
4. Colonna, Paul C.: Capsular arthroplasty for congenital dislocation of the hip, J. Bone & Joint Surg. 35A:179-197, 1952.
5. Crego, C. H., and Schwartzman, J. R.: Follow-up study of the early treatment of congenital dislocation of the hip, J. Bone Joint Surg. 30A:428-442, 1948.
6. Dickson, F. D.: Shelf procedure in congenital dislocation of the hip, J. Bone & Joint Surg. 17:43-47, 1935.
7. Dunlap, K., Shands, A. R., Hollister, L. C., Gaul, J. S., and Streit, H. A.: A new method for determination of torsion of the femur, J. Bone & Joint Surg. 35A: 289-311, 1953.
8. Faber, A.: Untersuchungen über die Aetiologie und Pathogenese der angeborenen Hüftverrenkung, Leipzig, Thieme, 1938.
9. Farrell, B. P., Lackum, von W. H., and Smith, A.: Congenital dislocation of the hip, J. Bone & Joint Surg. 8:551-556, 1926.
10. Galloway, Herbert P. H : Congenital dislocation of the hip, J. Bone & Joint Surg. 8:539-550, 1926.
11. Gardner, Ernest: Development of the Joints. Am Acad Orthop. Surgeons Instructional Course Lectures 9:149, 1952.
12. Gill, Bruce: End results of early treatment of congenital dislocation of the hip, J. Bone & Joint Surg. 30A:442-453, 1948.
13. Hart, V.: Primary genetic dysplasia of the hip with and without classical dislocation, J. Bone & Joint Surg. 24:753-771, 1942.
14. Heyman, C. H.: Congenital dislocation of the hip, J.A.M.A., 106:11-16, 1936.
15. Hibbs, R A Anteversion of the neck of the femur, J A M.A. 65:1801-1802, 1915.
16. Howorth, M B Congenital dislocation of the hip, Ann Surg 125:216-236, 1947.
17. ———: Shelf stabilization of the hip, J. Bone & Joint Surg. 17:945-952, 1935.
18. Inman, Verne· Functional aspects of the abductor muscles of the hip, J. Bone & Joint Surg. 29:607-619, 1947.
19. Jones, Robert, and Lovett, Robert W.: Orthopedic Surgery, p. 552, Baltimore, Wood, 1923.
20. Judet, J., Judet, R., Langrange, J., and Dunoyer, J.: A study of the arterial vascularization of the femoral neck in the adult, J. Bone & Joint Surg. 37A:663-680, 1955.
21. Leffman, Rudolph, and Pauker, Emil: Atraumatic treatment of congenital dislocation of the hip joint, J. Bone & Joint Surg. 36A:757-764, 1954.
22. McCarroll, H. R.: Early management of congenital dislocation of the hip *in* Regional Orthopaedic Surgery and Fundamental Orthopaedic Problems #2, pp. 125-156, Ann Arbor, Edwards, 1948.
23. Martz, Carl D., and Taylor, Clifford G.: The 45° angle roentgenographic study of the pelvis in congenital dislocation of the hip, J. Bone & Joint Surg. 36A:528-538, 1954.
24. Massie, William K.: Vascular epiphyseal changes in congenital dislocation of the hip, J. Bone & Joint Surg. 33A:284-306, 1951.
25. Massie, William K., and Howorth, M. Beckett: Congenital dislocation of the hip, J. Bone & Joint Surg. 32A:519-531, 1950.
26. ———: Congenital dislocation of the hip, J. Bone & Joint Surg. 33A:171-198, 1951.
27. Morrison, L. B.: A study of the hip joint from the standpoint of the roentgenologist, Am. J. Roentgenol. 28:484-520, 1932.
28. Ortolani, Marino: Congenital Dislocation of the Hip (New Diagnostic and Prophylactive Corrective Criteria), Istituto Provinerale per l'Infanzia, Center for the Diagnosis and Prophylaxis of Congenital Dislocation of the Hip, Ferrara, Italy.
29. Putti, Vittorio: Early treatment of congenital dislocation of the hip, J. Bone & Joint Surg. 11:789-809, 1929.
30. ———: Early treatment of congenital dislocation of the hip, J. Bone & Joint Surg. 16:13-21, 1933.
31. ———: Congenital dislocation of the hip, end results of closed reduction, Chir. org. movimento 20:93-112, 1934.
32. Ryder, Charles T., and Crane, Lawrence: Measuring femoral anteversion: the problem and a method, J Bone & Joint Surg. 35A:321-328, 1953.

33. Scaglietti, O.: Indirizzi odierni nel trattamento della lussazione congenita dell' anca, Chir. org. movimento 25:308-320, 1940.
34. Severin, Erik: Congenital dislocation of the hip joint (late results of closed reduction), Acta chir. scandinav., Supp. 63, 1941.
35. Strayer, L. M., Jr.: The embryology of the human hip joint, Yale J. Biol. & Med. 16:13-26, 1943.
36. Trueta, J., and Harrison, M. H. M.: The normal vascular anatomy of the femoral head in adult man, J. Bone & Joint Surg. 35B:442-461, 1953.
37. Tucker, F. R.: Arterial supply to the femoral head and its clinical importance, J. Bone & Joint Surg. 31B:82-93, 1949.
38. Wiberg, Gunnar: Studie dysplastic acetabula and congenital subluxation of the hip joint, Acta chir. scandinav., Supp. 58, 1939.
39. Wolcott, W. Eugene: The evolution of the circulation in the developing femoral head and neck, Surg., Gynec. & Obst. 77:61-68, 1943.

Dislocation Congenite del Coxa — Su Causas e su Effectos

Summario in Interlingua

Dislocation congenite del coxa resulta ab tensiones mechanic applicate a un articulation in que il existe un relaxation pathologic del capsula articular. Omne le altere alterationes pathologic—i.e. oblique tecto acetabular, deformitates del capite femoral, torsion femoral, e coxa valge—es responsas secundari effectuate per alterate tensiones mechanic. Le alterationes vascular que es communmente incontrate in le epiphyse post le termination del tractamento resulta ab un relativemente insufficiente arrangiamento vascular de character individual in un coxa specific. Iste arrangiamento augmenta le susceptibilitate del coxa a trauma de formas mesmo levissime. Le alterationes es comparabile al alterationes trovate in coxa plan in patientes de etates plus avantiate. Si non tractate, illos produce un grave deformitate al etate adulte. Con le exception de iste alterationes vascular, omne deformitates es revertibile e tractabile, sed le difficultates del tractamento accresce rapidemente post le secunde anno del vita del patiente e deveni si formidabile post le septime anno que le tractamento de un condition bilatere post iste etate non es recommendabile. Le tractamento durante le prime annos del vita consiste in le mantenentia de abduction con o sin rotation interne per le medios que es le melio cognoscite al chirurgo usque le relation normal inter capite e acetabulo es stabile. A etates plus avantiate le tractamento deveni multo individualisate, sed il ha bon rationes pro recommendar traction skeletal como mesura preparatori al technicas de reduction tanto aperte como etiam claudite. Le tractamento a iste etate e a etates ancora plus avantiatedeberea esser interprendite solmente per chirurgos qui possede le requirite fundo considerabile de experientias o sub le surveliantia directe de tal chirurgos, proque le integre futuro del coxa es in joco.

Si alterationes vascular pote esser evitate per extreme attention, omne coxa con dislocation congenite que es presentate ante le secunde anno del patiente pote esser subjicite a un tractamento que se termina con le attingimento de un coxa quasi normal ab le punctos de vista tanto anatomic como etiam functional, e omne coxa que es presentate a un etate plus avantiate va responder al reduction anatomic per un bon resultato permanente, providite que le reduction pote esser mantenite a omne tempores.

11

Persistent Hereditary Edema of the Legs —Milroy's Disease

JAMES HARVEY JENNETT, M.D.*

One of the unsolved and unsatisfactorily explained clinical entities in medicine is presented—a family in which for 6 generations of direct descent some members have been afflicted with hereditary edema of the legs.

I first saw members of this family in 1930 when Carl G., aged 14 (Fig. 1) came to the outpatient clinic at the Kansas City General Hospital with a painful ingrown toenail. During the examination it was noted that he had very large indurated edematous legs with excessive moisture about his feet and verruca accuminata scattered extensively over his toes. His legs had an elephantiasis-like appearance, the skin being tough and leathery, the hair follicles and the sweat glands markedly farther apart than normal, and the hard, brawny, white edema extended to the knees of both legs. In taking a history I found that his mother also had this abnormality, that a sister had the same trouble, and that all of them said they had been afflicted since birth.

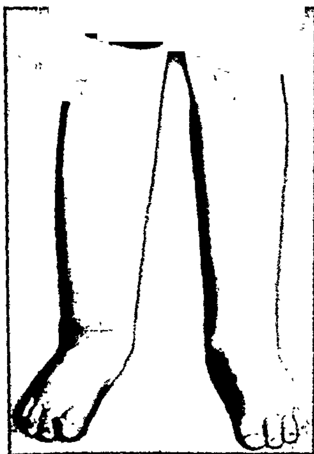


FIGURE 1
FIGS. 1-5, 11 & 12 (Jennett, J. H.: J Missouri M. A. 28:601-605)

Ernestine, Carl's sister, (Figs. 2 and 3) then aged 24, also had marked edema of both legs extending to the knees. She and her mother stated that they believed that she was born with bilateral clubfeet and that she always had had a spinal curvature. She did not learn to talk until she was 5 years old and always had had a speech defect resembling cleft palate, but examination revealed only a very arched deformity in the posterior portion of her hard palate. She began menstruating at age 13 but never has menstruated more than once or twice a year.

The mother, Hattie G. (Figs. 4 and 5), then aged 50, weighed 280 pounds and was 5 feet 10 inches tall. She made a very unusual appearance with the peculiar swinging gait caused by the weight of the heavy

*Kansas City, Mo.



FIGURE 2

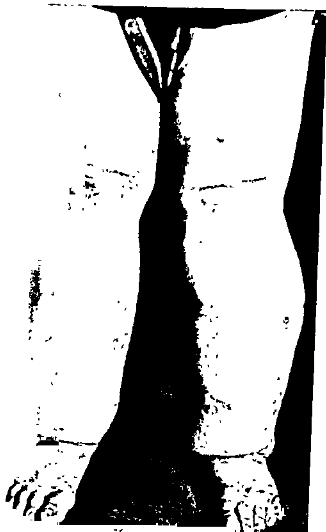


FIGURE 4



FIGURE 3



FIGURE 5

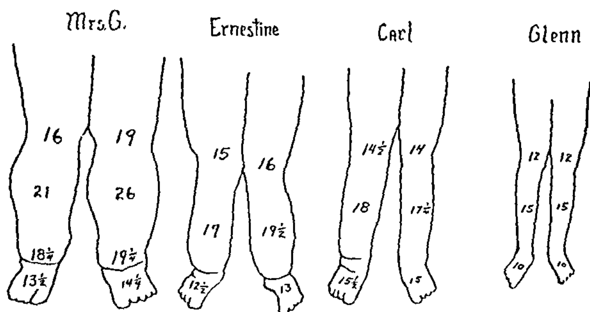


FIG. 6. (Measurements are given in inches.)

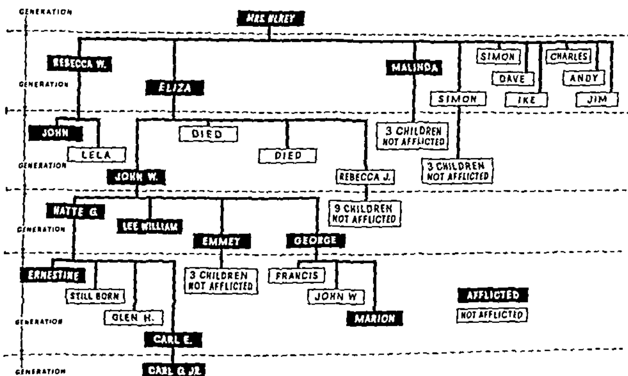


FIGURE 7

feet and legs beneath her and the long garments she wore to conceal her legs and feet. Beginning just above her shoetops there was a tremendous edema which extended to the knee in the right leg and to the midthigh in the left. The shape of her legs was very striking and seemed to be typical of this condition. There were constricting bands at the ankles, and the edematous tissue hung in folds over the shoetops—as Meige[4] says, "like the pants of a Zouave."

Another son, Glenn, then aged 19, was not afflicted with any swelling of his legs and seemed to be a perfectly normal boy except that he had a right inguinal hernia.

The circumference leg measurements of these patients is shown (Fig. 6).

The mother stated that in addition to these

3 children, her second child was stillborn and was not fully formed. The mother stated that during these 4 pregnancies her edematous legs were improved rather than made worse and that none of the 4 childbirths was prolonged or difficult.

With the mother's very co-operative assistance in searching family records and writing to relatives, the family record of those afflicted and those not afflicted is presented (Fig. 7). In 1955 Carl E. brought his son, Carl, Jr., to see me, stating that he also had been afflicted since birth. This 6th generation member, Carl, Jr., aged 16, is shown in Figures 8 and 9, on the left, standing by his father, now aged 40. Figure 7 shows 14 afflicted individuals in 6 generations among the 33 members of the family of which Mrs. G. can find record. The family ancestors of Mrs. G. lived in rural Missouri except for the first generation case shown in Figure 7, whom she says was a Pennsylvanian of German descent. In this family tree it is interesting to note that an unaffected parent never transmits the affliction. About as many males as females are affected. All have swelling in both legs except 2, George G. and Marion, who have it in the right foot and leg only. It may be transmitted from father to child or mother to child. There seems to be no interference with a long active life.

A general physical examination of these patients revealed no abnormality of the heart, the kidneys, the lungs or other organs or functions. The reflexes were present and active in all. Blood pressure, blood chemistry, blood count, Wassermann test, Kahn test, urine examination and other routine tests revealed nothing abnormal. An examination of the urine for chyluria was made on several occasions, but it was not found. Roentgenograms of the legs revealed no abnormality of the bones but did show the tremendous increase in the soft tissues with considerable irregular dense mottling scattered throughout. This mottling in the mother's legs has the density of calcium

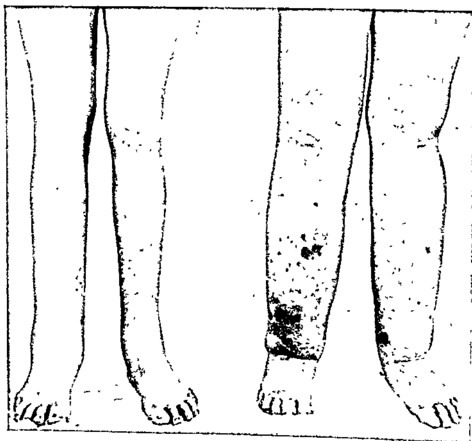


FIGURE 8

deposits in places. Plates of the lumbosacral spine were made in these 3 cases in 1930, and spondylolisthesis was ruled out in all. Ernestine had a marked scoliosis to the right in the lumbar region. Carl E. has a very suspicious spina bifida occulta, and Hattie G. has very markedly enlarged lateral spines to the lower lumbar vertebra. Roentgenograms of the sella turcica failed to show enough variation from the normal to suggest any abnormality of the pituitary gland. The mother reported that Ernestine had been given pituitary extract by a doctor for some time, previous to 1930, but it made her feel so miserable that she had had to discontinue it.

The basal metabolic rate was taken on these 3 cases in 1930 and repeated on each case, and all were found to be between minus 32 and minus 3. Because of this, thyroid was prescribed in reasonably adequate doses for 2 months for all 3 patients. It was discontinued because no benefit was noted.

Blood calcium was taken on each of these 3 patients in 1930. Ernestine had a blood serum calcium of 12.7; Carl, 10.5; and Hattie G., 11.2.

Biopsy specimens were taken from Ernestine, Carl E. and Hattie G. (Fig. 10). When the skin was incised it gapped widely, and watery lymph exuded freely until a small blood vessel was cut, and then bright red blood and lymphlike fluid ran freely. Healing took place quickly, and there seemed to be good resistance against infection. Under the skin was a very thick layer of fatty, collagenous or jellylike tissue, giving the large swollen appearance and doughy feel to the legs. Microscopic sections showed marked flattening of the dermal layers, excess fibrous tissue and collagen, all separated by intracellular spaces filled with edema. There was no lymphangiectasis, the lymphatics being hard to make out and not dilated. There is some perivascular infiltration and in places almost obliterative endarteritis of some of the small vessels.

The skin in these cases was not stretched tight and glossy as in dropsical edema. It was pachydermic in appearance and texture, could not be pinched up between the fingers,

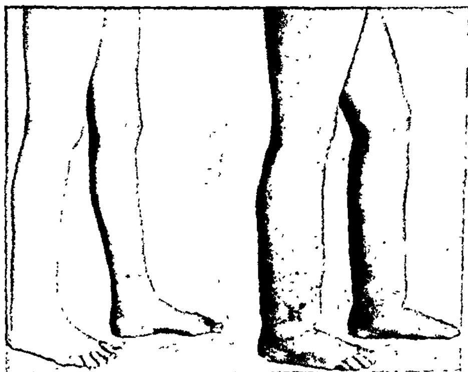


FIGURE 9

it blanched white on light pressure and yielded on prolonged hard pressure to form a deep pitting which would take from 10 to 15 minutes to fill out.

This condition never seemed to cause any pain or discomfort other than the weight and the clumsiness of the legs and the cosmetic embarrassment. The afflicted members of this family always have done hard work and been in good health. The children were able to keep up with their classmates in school. However, there have been acute attacks and remissions. The four of these patients whom I have seen on occasions during these 26 years have had occasional acute episodes of increased swelling of one or both legs, fever and chills, and have been acutely ill in bed for from 3 to 10 days. Carl, Sr., has had several such episodes but may go 10 years without one. I saw only one of these patients in such acute illness: Carl, Sr. It seemed to me to be a fungus and secondary infection of his feet with acute deep lymphangitis of both feet and legs. Carl, Jr., who was photographed in January, 1955, for this chapter (Figs. 8 & 9) then had very slight involvement in the left leg. His father reports that in May, 1956, Carl, Jr., had an acute illness with increased leg swelling and acute inflammation, with chills and fever to 107° F., and nearly died. He recovered completely from the acute illness, but, ever since, his left leg has been involved with the chronic edema to practically the same degree as his right. Hattie G. reports that on occasions she has noticed temporary sudden reduction in swelling with one stocking falling down because the swelling in that one leg seemed to be partially relieved temporarily. On other occasions after long hard work on her feet, she has stated that her left calf has swollen to 30 inches in circumference and that her feet and shoes became sopping wet with perspiration or other moisture ooze and that she had to crawl to the house on her hands and knees because her legs were so heavy.



FIGURE 10

They all report that if they stay off their feet the swelling is reduced. If they go to bed for a couple of weeks or keep the legs bound tightly day and night, the swelling will almost disappear, but it quickly returns when they get up. When the legs are kept firmly bound and the swelling is kept out of the legs they feel much lighter on their feet, but there is some swelling of the face and puffiness of the hands. They feel drowsy and sluggish. Because of this, the difficulty in keeping the legs bound and the chafing it causes on the legs, they all discontinued binding attempts.

There is no anesthesia of the skin. If any difference, their legs seem to be a little more sensitive than normal. The swelling is less, and they feel better on cold damp days than on warm days. A hot bath always increases the swelling.

Mrs. Hattie G. made some light-weight canvas leggings at my request (Fig. 11). At left the leggings are shown laced firmly on her legs with a 3-inch tongue down the middle the first day that they were applied. After she wore them day and night, lacing them snugly each morning, the center illustration shows them laced together. The illustration at right shows that in addition to their coming together in front a 1-inch tuck was taken up the entire back, showing

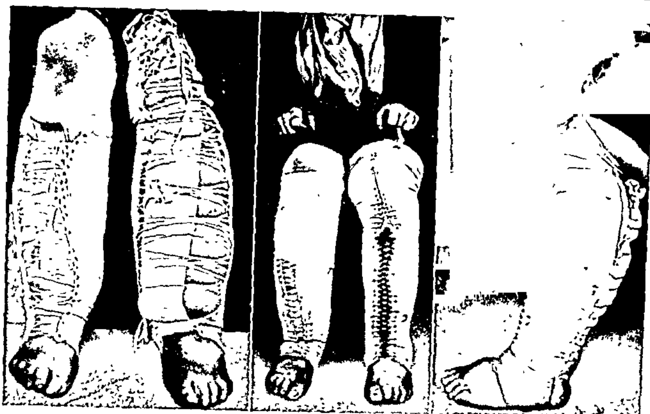


FIGURE 11

the degree of fluid that can be expressed from her legs with simple constant pressure. Figure 12 shows Ernestine and Carl E. wearing high laced boots which they could hardly get on at first, but after one week the boots could be laced together. These boots gave them great joy for the first few days but they soon discarded them as not worth the effort.

In 1931 I asked a surgeon friend to do the Sistrunk[10] modification of the Kondoleon[3] operation on the lateral aspect of both of Ernestine's legs (Fig. 13). Long strips of tissue, including everything from skin to muscle surface, from knee to external malleolus, 2 inches wide, were removed. Later observation for 2 years and her opinion regarding the result were not very encouraging. There were no unfavorable aftereffects, but there was hardly enough improvement to have made it worth while.

So far no other treatment has produced beneficial results. We have not tried any sympathectomy or ligation procedures. A suggested form of treatment that I have not had the opportunity to try on my patients is to have them sleep on a Sanders' rocking bed for possible daily temporary reduction of leg size and weight.

In 1892, Milroy,[5] of Omaha, Neb., described the unusual condition so typically illustrated by this family, to which Osler[7] gave the name Milroy disease. In Milroy's series there were 22 cases among 97 individuals in 6 generations of one family. Our cases resemble Milroy's exactly in that (1) there is a strong familial predisposition; (2) it begins at birth; (3) males and females are affected equally; (4) the edema is confined to the legs; (5) there is no pain or discomfort except weight of the legs; (6) there is no demonstrable cause, local or general; (7) there is an absence of constitutional symptoms; (8) it is not inimical to life; (9) no treatment gives more than temporary relief; (10) it never is cured. To this I would add (11) that it never skips a generation, and an unafflicted member of the family

never transmits it, and (12) that afflicted individuals may or may not transmit it.

Relatively few families with Milroy's disease have been described in the medical literature. No one ever has described as large a series as Milroy's or traced it in a family so extensively. Nonne,[6] of Germany, gave the first published description of such similar condition in 1891. In 1898 Meige,[4] in France, presented 8 similar cases in 4 generations, but in all of his cases the edema did not begin until puberty. In 1902 Rolleston,[9] in England, presented 3 cases in 2 generations. In 1908 Hope and French,[2] in England, reported 13 cases in 5 generations, but practically all of them had questionable features. From 1908 until 1930 very little was written about this curious affliction. Since then much has been written about lymphedema, and many scattered cases of

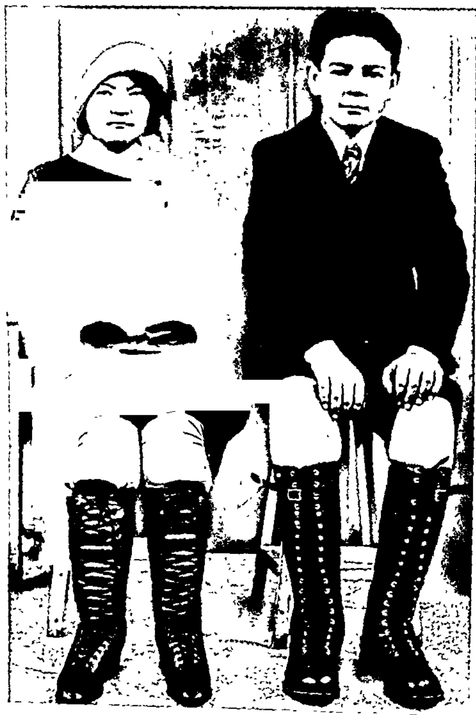


FIGURE 12

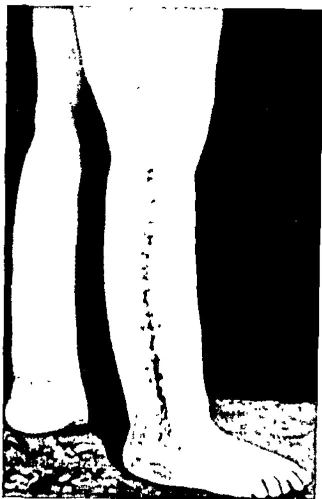


Figure 13

similar affliction, some congenital, some with or without heredity record, have been described.

The etiology of this curious affliction is still obscure. So far as I have been able to determine there never has been an autopsy reported on one of these cases. There have been many theories of explanation of this condition. Milroy suggested that possibly there was a congenital absence of valves in the large veins of the legs. I never have been able to determine or find whether or not this has been adequately ruled out as a possibility. Osler gave it as his opinion that this disease was an angioneurotic type of edema. I believe this was because he saw acute exacerbations of fungus and secondary infection superimposed on chronic lymphedema. Hope and French considered it to be a vasomotor neurosis.

Could there be some congenital anatomic abnormality, the nature of which has not been explained? Apparently there is a local rather than a generalized cause, and it is an obstruction of some kind.

Elephantiasis from filarial infection, as seen in natives of Africa and India, is considered to be a lymphatic obstruction. Milroy's disease could be a similar lymphatic obstruction from fibrosing, swelling or plugging of lymphatics from fungal or bacterial infections of the feet and carried up the lymphatics in early childhood or later. The hereditary feature could be a hereditary lack of immunity or a hereditary hypersensitivity to such infection. This reasoning would be unacceptable if these patients were born with this condition or had it since birth. However, in questioning the mothers among my patients with this trouble, the information is exceedingly indefinite as to the age at which the defect first was noticed. I could not obtain any pictures of these afflicted individuals in their infancy. The earliest age at which I saw any of these cases was 14 years. Seeing the leg size of my patients grow larger in proportion as they grow older, get worse after acute episodes of chills and fever, and *seeing Carl, Jr., with only slight involvement* in the left leg up until the spring of 1955, but now with the left leg practically as large as the right since his severe acute episode with fever of 107°F. in the spring of 1956, it seems to me that this theory of infection and possible hereditary lack of immunity or a hereditary hypersensitivity could be the etiologic explanation.

A recent paper[8] described "dramatic" improvement in a case of Milroy's disease *treated with prednisone*, one of the latest steroids of the cortisone type. On reading this I started Carl, Jr., on Co-Hydra-Deltra (prednisolone) by mouth, 5 mg. 4 times a day, and Carl, Sr., on Meticorten (prednisone), 10 mg. 4 times a day. *I measured* their calves and ankles before and after 10 days of this heavy dosage and saw no

improvement whatsoever. These 2 patients see no improvement in their condition or the way they feel.

This limited experience with this treatment of my 2 cases and the "dramatic" improvement in the single patient referred to in the recent paper [8] are inadequate for any worth-while conclusion. The "dramatic" improvement could have been attributable to the elastic stockings and the postural exercises that also were prescribed. Ernestine and Carl, Sr., thought their improvement "dramatic" for a while when in 1930 they wore the high boots shown in Figure 12. I do not believe that the steroids hold forth any real hope of benefit in this affliction.

For excellent presentation of the general subject of lymphedema with more recent bibliography reference list, see reference 1.

REFERENCES

1. Allen, E. V., Barker, N. W., and Hines, E. A.: Peripheral Vascular Diseases, ed. 2, Philadelphia, Saunders, 1955.
2. Hope, W. B., and French, Herbert: Quart. J. Med. Oxford 1907-8, i, pp. 312-330.
3. Kondoleon, E.: München. med. Wchnschr. 59:525, 1912.
4. Meige, Henry: N. Iconog. de la Salpêtrière 14:453, 1899.
5. Milroy, W. F.: New York J. Med. 56: 505, 1892.
6. Nonne, M.: Virchows Arch. path. Anat. 125:189, 1891.
7. Osler, William, and McCrae, Thomas: *Principles and Practice of Medicine*, p. 1149, New York, Appleton, 1927.
8. Panos, T. C.: J.A.M.A. 161:1475, 1956.
9. Rolleston, H. D.: Lancet 2:805, 1902.
10. Sistrunk, W. E.: Ann. Surg. 85:190-193, 1927.

Persistente Edema Hereditari del Gambas—Morbo de Milroy

Summario in Interlingua

Es presentate le historia de un serie de casos de persistente edema hereditari e congenite del gambas, occurrente in sex generationes de descendita directe de un familia de Missouri in que dece-quatro ex trenta-tres membros esseva affligite. Es includite un representation graphic del arbore genealogic e numerose photographias. Iste casos satisface le mesme criterios que le casos del serie de Milroy pro le quales Osler ha introducite le nomine morbo de Milroy. Es signalate le absentia (usque nunc) de un adequate explication technic. Extense studios del casos presente revelava nihil promittente ab le puncto de vista del explication diagnostic. Le affligite membros de iste familia exhibi multe altere defectos congenite. Le acute episodios que es interpretate per certe autores como de natura angioneurotic o como neurosis representa in le opinion del autor lymphangitis a infection fungal e secundari. Es describite e commentate le tentativa de bandagear le gambas e le effectos posterior de un typo de operation Sistrunk o Kondoleon executate in un caso.

12

Osteogenesis Imperfecta: Some Clinical and Genetic Considerations

C. NASH HERNDON, M.D.*

TERMINOLOGY

Osteogenesis imperfecta has been defined as a generalized inherited mesenchymal defect characterized by abnormal fragility of bone, a bluish appearance of the sclerae, and frequent development of otosclerosis. Unfortunately, this condition has been assigned many different names, and more than 20 synonyms may be noted in the literature of recent years. While the term osteogenesis imperfecta seems to be most firmly entrenched in the English literature, the term *fragilitas ossium* is recommended by the American Medical Association for diagnosis coding of hospital records (Plunkett & Hayden, 1952). The term osteopsathyrosis also has some currency, and the eponyms in use include Lobstein's disease, van der Hoeve's syndrome, Eddowe's syndrome and Vrolik's disease. Each of these eponyms is used in a rather restricted sense to describe a particular combination of symptoms. As some cases are clinically apparent at birth and others do not exhibit pronounced symptoms until later in life, it has been customary to distinguish these groups as *osteogenesis imperfecta congenita* and *osteogenesis imperfecta tarda.* Seedorff's monograph (1949) discusses terminology, and gives a careful historic survey with a bibliography of 369 references

* Professor of Medical Genetics, Bowman Gray School of Medicine of Wake Forest College, Winston-Salem, N C

CLINICAL ASPECTS

The clinical manifestations of osteogenesis imperfecta are quite variable. Recently McKusick (1956) has given an excellent summary of the clinical findings, emphasizing the fact that the disease "has wide systemic manifestations and an exceedingly great range of clinical severity." The most outstanding symptoms and signs are associated with the skeletal system. Pathologic and histochemical studies have shown that while the growth and development of cartilage is normal, there is a derangement in the formation of osteoid, the organic matrix of bone, probably secondary to some defect of the osteoblasts (Follis, 1953). Biophysical studies indicate that there is an abnormal distribution of mineral salts and arrangement of organic fibers, with the bone remaining largely immature and failing to undergo the usual transformation to compact bone (Engfeldt *et al.,* 1954). Often there is deficiency in growth in length of the long bones, due to irregular development at the epiphyseal plate. These defects of bone formation and structure result in abnormal bone fragility. Thus the occurrence of fractures as a result of trivial trauma is the most prominent feature of the disease. These fractures usually heal well, but the formation of excessive amounts of callus, sometimes mistaken for osteosarcoma (Strach, 1953), and the development of pseudoarthroses are occasional complications. Mal-

union with angulation is often observed. As the bones are also more plastic than normal, bowing of long bones is not infrequent, an anterior bowing of the tibia being seen most often. In severe cases the long bones may be markedly shortened, with micromelia being one of the characteristics of the congenital type. Various malformations of the spine and the thoracic cage may occur. The skull frequently presents platybasia and frontal bossing, and often numerous wormian bones are found in the occipital region similar to those seen in cleidocranial dysostosis. The teeth are quite susceptible to caries and often present a yellowish-brown discoloration apparently secondary to defective dentine formation (Roberts & Schour, 1939). The tendons and the joint capsules are also poorly developed, resulting in laxness of ligaments and excessive mobility of joints.

Often the blue color of the sclerae has been emphasized as an important diagnostic sign. This is described as a milky blue, slate blue or robin's-egg blue. In many patients this color is quite pronounced, in others it may be inconspicuous, and occasionally it is absent in unquestioned cases of the disease. The blue color is due to increased translucency of the sclera, which permits the color of the underlying choroidal venous plexus to become visible. Pathologic studies of the eye have demonstrated a general deficiency of collagen and retention of immature precollagenous fibers (Ruedemann, 1953). Other ocular defects presumably based on the same connective tissue deficiency are reported occasionally, including embryotoxon, keratoconus, megalocornea and glaucoma.

The deafness which frequently develops during the third decade is clinically indistinguishable from otosclerosis but must be regarded as a special type that is etiologically and histologically distinct. Sclerosis of the petrous portion of the temporal bone may be demonstrated by roentgenograms. Ankylosis of the stapes is the usual lesion, although the cochlear type is encountered occasionally. The patient may experience tinnitus or vertigo. Pregnancy and middle ear infections may precipitate or aggravate the symptoms of otosclerosis. Pregnancy is also a hazard from the viewpoint of complications in delivery, and pelvic deformities may necessitate cesarean section (Staples & Riva, 1954).

Other manifestations secondary to the widespread defect of collagenous tissue are encountered occasionally. Neurologic signs, secondary to pressure on the cranial nerves or the spinal cord at the level of the foramen magnum may occur with the more severe degrees of platybasia. The skin is often thin and atrophic in appearance, with a tendency to hypertrophic scar tissue formation (Scott & Stiris, 1953).

As McKusick (1956) and others have emphasized repeatedly, the range of clinical severity in osteogenesis imperfecta is extreme. In severe cases bony development is quite deficient, multiple fractures may occur in utero, and the affected infant may be stillborn with tiny extremities and caput membranaceum. Some infants may sustain fractures in the process of delivery, while in others fractures may not appear until the child attempts to walk. In milder cases fractures may be few in number and appear only later in life. Some individuals never develop fractures but may be identified as affected by blueness of the sclerae or minor changes noticeable in roentgenograms. The blue color of the sclerae is apparently the most constant feature of the disease in affected families, but even this may be absent in some individuals. From a study of case reports in the literature, Bell (1928) concludes that among adults with blue sclerae, about 60 per cent have a tendency to fractures, about 60 per cent have otosclerosis, and about 44 per cent have all 3 defects. There is genetic evidence, discussed below, to suggest that some cases are subclinical and may be mild enough to escape detection.

INHERITANCE

Osteogenesis imperfecta has been recognized as an inherited condition for many years. Seedorff (1949) gives a translation of a description of this disease in 3 generations of a family by Ekman in 1788. Since then numerous pedigrees and case reports of individual families and several careful studies of a series of cases have appeared. There is overwhelming evidence that osteogenesis imperfecta is due usually, and possibly always, to an autosomal dominant gene. This genetic mechanism implies that any affected person, if married to a normal individual, will transmit the disease to approximately half of the children. It is well known that the chromosomes, and the genes located in the chromosomes, occur in pairs. The autosomal dominant mechanism of inheritance postulates that the affected individual possesses one pathologic or mutant gene paired with a normal gene, located on one of the 22 pairs of chromosomes other than the X and the Y or sex chromosomes. A mutant gene is classified as dominant if its physiologic effect is stronger than that of the normal gene with which it is paired, its allele. Therefore, a dominant mutant gene usually will produce a recognizable abnormality even when paired with a normal gene. A person having one mutant and one normal gene paired is said to be heterozygous for the mutant gene, while an individual with two identical mutant genes paired is said to be homozygous for the mutant gene. Patients with osteogenesis imperfecta are thus thought to be heterozygous for the mutant gene responsible for this disease. An individual heterozygous for a mutant gene will form 2 kinds of gametes (sperm or ova), half containing the mutant gene and half containing the normal allele. If the spouse is producing only normal gametes, then half of the children would also be heterozygous for the mutant gene The exact physiologic effect of the mutant gene is unknown, but it apparently produces a generalized mesenchymal defect which inhibits the maturation of collagen beyond the reticulin fiber stage (McKusick, 1956).

A large number of extensive pedigrees have been published demonstrating typical dominant transmission of the full syndrome of osteogenesis imperfecta through several generations (Bell, 1928). As many as 27 affected persons in 5 generations of one family have been described (Hills and McLanahan, 1937). Such pedigrees are in full agreement with the mechanism described previously. On the other hand, there are also numerous observations which at first sight do not seem to be compatible with the dominant gene theory. These observations generally fall into 2 groups: (1) apparently irregular pedigrees, in which an occasional normal person will transmit the disease to one or more children; (2) apparently sporadic cases, with a family history entirely negative for osteogenesis imperfecta. These situations must be accounted for adequately if the theory of dominant transmission is to be considered as established.

Irregular pedigrees, which may be disturbing at first sight, are readily explained by appeal to a principle which has been shown to have wide applicability in human genetics, that of reduced penetrance. The concept of reduced penetrance of genes is simply the geneticist's way of expressing the common clinical observation that there may be subclinical cases of a disease that are not diagnosed. While the concept of the subclinical case is general knowledge in connection with infectious diseases, it is not mentioned so often in connection with constitutional disease. With regard to inherited disease, it must be remembered that the presence of an abnormal gene in a fertilized ovum is not equivalent to the development of disease in the individual. A number of intermediate steps, involving biochemical, physiologic and physical factors, must occur. While a pathologic gene may initiate a given series of reactions or may set limits within which certain reactions must occur, this series of reactions might be modified by a

OSTEOGENESIS IMPERFECTA

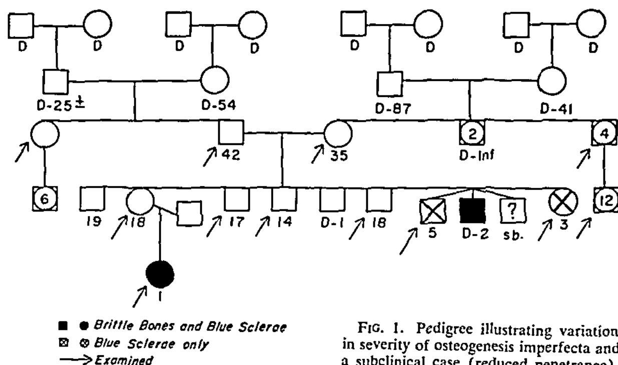


FIG. 1. Pedigree illustrating variation in severity of osteogenesis imperfecta and a subclinical case (reduced penetrance).

number of possible factors. It is easily conceivable that one individual possessing a pathologic gene might react by developing a clinically severe abnormality, while another individual with the same gene might be able to compensate for the abnormality partially or completely and thus present a very mild case, a forme fruste, or a subclinical and asymptomatic form of the condition. The recognition of subclinical cases may depend on the level of diagnostic skill or the availability of specific laboratory tests at a given time. It must be admitted that the fact that a given individual may represent an unrecognized and asymptomatic case does not alter his genetic composition or genotype, and that the subclinical carrier of genetic disease is fully capable of transmitting the defect to his offspring.

An example of reduced penetrance or a subclinical case is seen in Figure 1. This is the pedigree of a family taken from a series of 33 families with osteogenesis imperfecta studied by the Department of Medical Genetics of the Bowman Gray School of Medicine from 1940 to 1955. In the central sibship (4th generation) of Figure 1, there were 10 children. The first 6 were normal with regard to bone fragility, although one died of diphtheria at age 1 year. The 7th pregnancy of the mother resulted in triplet boys, of whom one was a stillborn macerated fetus. Autopsy of the stillborn child was not obtained, and it is not known whether or not he presented any signs of osteogenesis imperfecta. On the day after birth the parents discovered a fracture of the leg of the second-born triplet. The child was referred to a hospital at the age of 1 month when roentgenograms demonstrated 7 fractures, involving 3 of the extremities and 2 ribs. All long bones were short and stubby with poor calcium content and with bowing deformities. Blood calcium, phosphorus and phosphatase studies were normal. This child continued to have fractures and increasing deformities and expired with bronchopneumonia shortly before the 2nd birthday. The surviving member of the triplet set was last examined at the age of 5. He presented a

typical china-blue color of the sclerae, which had been noted in infancy but had sustained no fractures. As it was not possible to obtain extensive blood typing and palm and finger print studies for diagnosis of zygosity, it is not known whether these triplets were derived from 1, 2 or 3 ova. A younger sister of the triplets, when last seen at the age of 3, also had china-blue sclerae but had sustained no fractures. The important part of this pedigree concerns the older sister of the triplets, aged 18 when last seen, who had a daughter 1 year old with the severe form of the disease. This child had blue sclerae and had sustained 5 fractures of the extremities. Roentgenograms showed typical bone changes of osteogenesis imperfecta. The mother of this child was entirely normal on physical examination and did not have blue sclerae. By roentgenogram the cortices of some long bones were questionably thin but were within the usually accepted normal range. The conclusion seems inescapable that this woman, with an affected child and 3 affected sibs, must represent a subclinical case not diagnosable by physical or radiologic examination. She must also be heterozygous for the dominant gene which produced brittle bones and blue sclerae in her daughter and one brother, and blue sclerae alone in another brother and sister. The parents of this woman were also examined and were normal, but it seems likely that one of them may also be a subclinical carrier. Examination of the uncles, the aunts and several first cousins of the triplets was also essentially negative

The existence of subclinical carriers of osteogenesis imperfecta has 2 important implications. From the strictly practical viewpoint, it is not safe to assure an apparently normal sib or child of a patient with this condition that their children will all escape the disease. The undetected carrier may also transmit the disease It is useful to express the penetrance of a gene as the frequency, usually stated as a percentage, with which the pathologic gene produces clinical disease. In these terms a disease due to a dominant gene with 90 per cent penetrance would produce clinically apparent disease in 90 per cent of persons having the gene, while 10 per cent would be subclinical carriers. If the penetrance figure is known with accuracy, one then has a risk figure for the appearance of the abnormality in the offspring of apparently normal sibs or children of patients. Unfortunately, we do not as yet have sufficient data to specify a penetrance rate for the gene responsible for osteogenesis imperfecta.

The second implication of the demonstration of reduced penetrance of the gene for osteogenesis imperfecta is a purely theoretical one but gives some hope for the development of a rational method of therapy. If some individuals possessing the abnormal gene have no clinical evidence of the disease, some mechanism must exist which is able to compensate for or to suppress the pathologic effect of this gene. It is at least possible that this mechanism, completely unknown at present, might be susceptible to deliberate induction or intelligent manipulation as a form of therapy. It is possible that further study of families with osteogenesis imperfecta, and particularly of genetically identified carriers, might yield some clue leading to identification of the postulated compensating mechanism.

The second observation mentioned earlier which must be explained if the theory of dominant gene etiology of osteogenesis imperfecta is considered as established is the frequent appearance of apparently sporadic cases of the disease. Sporadic cases may represent one or more of several possible genetic situations. First, it is entirely likely that certainly some and possibly all of the sporadic cases may represent the first clinical appearance of a new mutation in the family under consideration. It is well established that new mutations are constantly appearing in families in which they have not been known previously. Most of these are duplicates of mutations that have ap-

peared previously in other families, and most of them are also pathologic, producing hereditary disease, although some mutations are advantageous to the individual, and the gradual accumulation of advantageous mutations is one of the basic mechanisms of evolution. It appears that certain normal genes are relatively unstable and frequently undergo mutation to form specific pathologic genes, the rate of this transformation being termed the mutation rate of the pathologic gene, usually expressed in terms of chromosomes per generation.

An example of the appearance of a new mutation of the gene for osteogenesis imperfecta may be seen in Figure 2. This pedigree describes a family in which 3 generations of patients with osteogenesis imperfecta now exist. However, it will be noted that if this family had been studied 50 years ago, before the persons in the 2 youngest generations were born, it would have given the appearance of a sporadic case. At this time, the situation was that of one female patient with typical osteogenesis imperfecta with blue sclerae, with 10 normal sibs living, normal parents and grandparents, and no history of any similar defect in any known relative. Later events proved that the usual dominant gene was present in this family, as the affected woman had 2 children with osteogenesis imperfecta, and her affected son also has a son with the same condition. Also it should be noted that the 10 normal sibs of the original patient had only normal children and grandchildren. It is thus apparent that many of the sporadic cases seen are sporadic only because they have no affected children, and represent the first clinical appearance of a new mutation.

A second explanation for the appearance of sporadic cases is the possibility that some cases might be due to a different gene, either autosomal recessive or sex-linked. If

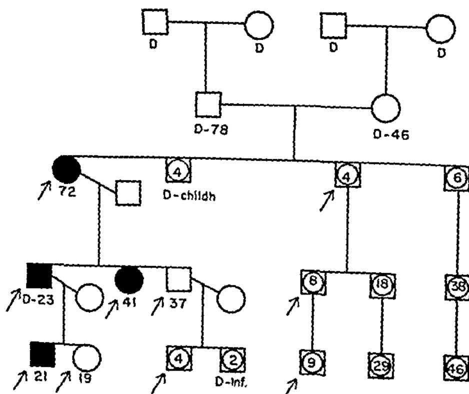


FIG. 2. Pedigree showing first clinical appearance of new mutation and its transmission to 2 additional generations.

a recessive gene were involved, the affected children would be homozygous for the pathologic gene, and the normal parents would be both heterozygous for this gene, or carriers. As only 25 per cent of the children of a marriage of 2 carriers would be expected to be homozygous, many families would contain only one affected child. As any postulated recessive gene for osteogenesis imperfecta must be rare in the general population, the mating of 2 unrelated carriers would be quite uncommon, and the marriage of cousins would provide a large proportion of the parents of affected children. Cousin marriage among the parents of patients with osteogenesis imperfecta has been reported only rarely, and there is no evidence that cousin marriage is of any significance in relation to this disease. While the possibility of recessive inheritance cannot be excluded on this basis, it at least becomes unlikely. If a proportion of sporadic cases were due to a sex-linked gene, we would expect to find more affected males than females in this group of patients. No disturbance of sex ratio among sporadic cases appears in systematically collected material. In the series of 180 cases in 55 families examined by Seedorf (1949), there are 33 sporadic cases, 15 of these male and 18 female. In the series of 72 cases in 33 families seen by the writer, 18 were sporadic, 8 were male and 10 female. There is no evidence to suggest that a sex-linked type of osteogenesis imperfecta might exist.

The third possibility suggested by the sporadic cases is that some of these might be due to purely environmental influences and not to genetic abnormality. In experimental animals it has been demonstrated that many genetic abnormalities can be duplicated by environmental shock (temperature change, chemicals, etc.) applied to the developing embryo at a critical stage of development. Goldschmidt (1955) has designated these environmentally produced mimics of hereditary disease as phenocopies. It seems certain that phenocopies also exist in man. For example, microcephaly is due to a recessive gene in some families, but it is also known that the condition may be produced by certain environmental agents (maternal rubella, radiation) acting early in pregnancy when the brain is developing rapidly. At present we have no evidence that any environmental change can produce the clinical picture of osteogenesis imperfecta, and it is difficult to conceive of any

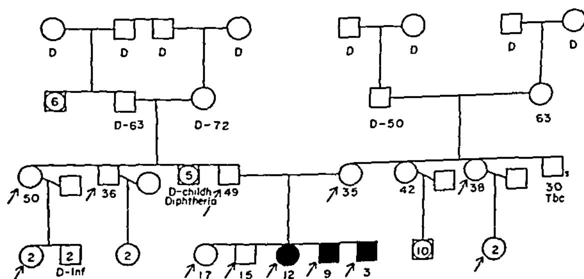


FIG. 3. Three cases of osteogenesis imperfecta in one sibship, other relatives normal.

combination of events compatible with life that could produce this specific and widespread damage to developing collagenous structures. While this possibility cannot be dismissed on the basis of presently available knowledge, it at least seems to be rather unlikely. In summary, it seems to be established that most, if not all, sporadic cases of osteogenesis imperfecta represent mutations of a dominant gene, but the possibility that some cases may be due to a recessive gene cannot be excluded.

The possibility that a genetically recessive type of osteogenesis imperfecta might exist is somewhat strengthened by the occasional observation of 2 or more cases in a single sibship, as is illustrated in Figure 3. In this family, 3 children presented typical osteogenesis imperfecta with deformities, recurring fractures and blue sclerae, while 2 sibs were normal on examination. Both parents, 3 uncles and aunts and 4 first cousins were normal on examination, and other relatives were reported to be normal. The paternal grandparents of the patients were first cousins, but this is not significant with regard to the affected children. This pedigree might easily be interpreted as suggesting the action of a recessive gene. Pedigrees of this type are apparently uncommon. McKusick (1956) remarks that he is not aware of the occurrence of 2 such offspring from parents who are indubitably normal, and that even if the parents were normal by every gauge, suspicion of subtle abnormality would remain. McKusick's suspicion of subtle abnormality is certainly well founded. It will be noted that the pedigrees in Figures 1 and 3 are distinguishable only because of the appearance of an affected child in the youngest generation in Figure 1, and it seems likely that the mechanism of inheritance is the same in both families. It seems more logical to suspect that one of the parents in Figure 3 might be a subclinical carrier of a dominant gene than to postulate that this family represents an etiologically distinct variety of osteogenesis imperfecta due to a recessive gene. Certainly there is no clinically apparent difference in the disease present in the family depicted in Figure 3 and that present in families with undoubted dominant transmission of osteogenesis imperfecta. However, the question of the possible existence of a recessive type must remain open until a sufficient number of systematically collected cases are accumulated to permit a statistically reliable decision. Individual case reports from the literature, which probably are biased in favor of reporting the unusual, are not reliable for this purpose. The largest series of cases yet reported, that of Seedorf (1949), is not sufficient to exclude the possibility of recessive inheritance and is also not sufficient to estimate either the mutation rate or the gene frequency of the known dominant gene with accuracy.

While the data are insufficient to estimate the mutation rate in absolute terms, it is of some interest to estimate the proportion of cases seen in practice that represent the first clinical appearance of a new mutation. Our own figures, based on 72 cases in 33 families, agree well with those of Seedorf (1949) based on 180 cases in 55 families, and these have been combined. In the total material, 51 cases were sporadic, with family history negative for similar affliction. If one considers this on the basis of family groups only, 58 per cent of the families seen contained only 1 recognized case. In terms of total cases, the 51 with negative family history would be 20 per cent of the total, but it must be remembered that not all of these cases were living at one time, and some were so mild that they would not present as patients. While an exact figure cannot be given, it is estimated that not less than one third of the cases seen by physicians would be expected to give a history negative for other cases in the family.

SUMMARY AND CONCLUSIONS

The clinical picture of osteogenesis imperfecta is summarized briefly, and refer-

ence is made to more extensive clinical discussions in the literature. The extreme range of clinical severity, from severe and often fatal cases present at birth to individuals identified only by blueness of the sclerae, is emphasized.

Evidence concerning the mechanism of inheritance of osteogenesis imperfecta is examined critically. Evidence is presented for the existence of subclinical cases or carriers, asymptomatic and not diagnosable by presently available technics. It is concluded that certainly the majority, and perhaps all, of the cases are due to an autosomal dominant gene which produces a generalized mesenchymal defect with inhibition of the maturation of collagen. The possibility that some cases may be due to an autosomal recessive gene cannot be excluded. Apparently sporadic cases, with family history negative for similar disease in any relative, are frequent, and most if not all of these represent the first clinical appearance of a new dominant mutation. It is estimated that about one third of the cases seen in practice will give no history of an affected relative.

REFERENCES

Bell, J.: Blue sclerotics and fragility of bone *in* Treasury of Human Inheritance, vol. 2, part 3, 1928.

Engfeldt, B., Engström, A., and Zetterström, R.: Biophysical studies of the bone tissue in osteogenesis imperfecta, J. Bone & Joint Surg. 36B:654-661, 1954.

Follis, R. H.: Histochemical studies on cartilage and bone: III. Osteogenesis imperfecta, Bull. Johns Hopkins Hosp. 93:386-391, 1953.

Goldschmidt, R. B.: Theoretical Genetics, pp. 255-266, Berkeley, Univ. Calif. Press, 1955.

Hills, R. G., and McLanahan, S.: Brittle bones and blue scleras in five generations, Arch. Int. Med. 59:41-55, 1937.

McKusick, V. A.: Heritable disorders of connective tissue: V. Osteogenesis imperfecta, J. Chronic Dis. 3:180-202, 1956.

Plunkett, R. J., and Hayden, A. C.: Standard Nomenclature of Diseases and Operations, ed. 4, Philadelphia, Blakiston, 1952.

Roberts, E., and Schour, I.: Hereditary opalescent dentine (dentinogenesis imperfecta), Am. J. Orthodontics 25:267-276, 1939.

Ruedemann, A. D.: Osteogenesis imperfecta congenita and blue sclerotics: a clinicopathologic study, A.M.A. Arch. Ophth. 49: 6-16, 1953.

Scott, D., and Stiris, G.: Osteogenesis imperfecta tarda: a study of 3 families with special reference to scar formation, Acta med. scandinav. 145:237-257, 1953.

Seedorff, K. S.: Osteogenesis imperfecta: a study of clinical features and heredity based on 55 Danish families comprising 180 affected members (Op. Dom. hered. hum., vol. 20), Copenhagen, Munksgaard, 1949.

Staples, P. P., and Riva, H. L.: Maternal osteogenesis imperfecta: report of 2 cases in sisters, Obst. & Gynec. 4:557-561, 1954.

Strach, E. H.: Hyperplastic callus formation in osteogenesis imperfecta: report of a case and review of the literature, J. Bone & Joint Surg 35B:417-422, 1953.

Osteogenese Imperfecte Alicun Considerationes Clinic e Genetic

Summario in Interlingua

Osteogenese imperfecte es un generalisate e hereditate defecto mesenchymal que es characterisate per un fragilitate anormal del ossos e resulta in recurrente fracturas e deformitates, un apparentia blauastre del sclera, e le frequente disveloppamento de otosclerosis Il existe in iste condition un extensissime scala de severitate clinic que varia ab grave e frequentemente mortal formas manifeste al tempore del nascentia usque a leve formas que es identificate solmente per le color blau del sclera.

Es presentate un examine critic de factos e observationes relative al mechanismo del hereditage de osteogenese imperfecte. Il ha observationes que indica que il existe portatores de iste condition in formas subclinic que non es directemente diagnosticabile per le nunc disponibile technicas. Iste portatores es ben capace a transmitter le morbo a lor

infantes, e le plus sever formas pote occurrer inter le infantes de portatores asymptomatic. Es formulate le conclusion que certo le majoritate del casos, e forsan omnes, es debite a un gen dominante autosomal que produce un generalisate defecto mesenchymal con inhibition del maturation collagenic. Le possibilitate que certe casos es debite a un gen recessive autosomal non pote esser rejicite completemente. Apparentemente, casos sporadic es frequente con historias familial que es completemente negative pro omne morbos affin. Le majoritate del casos sporadic, e possibilemente omnes, representa le prime manifestation clinic de un nove mutation dominante. Es a expectar que casos sporadic que attinge lor maturitate transmitte le morbo, como regula median, a levemente minus que un medietate de lor infantes. Nos estima que circa un tertio del casos presentate in le practica medical non ha un historia familial de occurrentias del morbo in consanguineos.

13

Heredity As a Factor in Malignancy

H. WINNETT ORR, M.D.*

When the occurrence of lung cancer became a very hot topic in both the medical and lay press, I returned to a study that I had made some years ago of heredity as a factor in malignant and other diseases. As a result of that investigation I had come to certain conclusions with reference, particularly, to the occurrence of cancer as shown in many generations of mice by Dr. Maude Slye,† in Chicago.

Dr. Slye found upon a study of thousands of mice, through many generations, that dominant, recessive and hybrid tendencies could be tabulated, recorded and reduced to statistics, and that definite conclusions could be arrived at, as to the occurrence, or the exclusion, of cancer from families of mice, and that for each selected series of breedings, predictions could be made as to the appearance or the nonappearance of cancer in succeeding families and individuals.

Dr. Slye said:

> In the human species, where pure strains are very rare, and where hybrids of unknown antecedents are the rule, it is practically impossible to know whether or not certain characteristics will appear.
>
> To a certain extent, however, and with a fair knowledge of our antecedents we may often be able to say that we do, or do not, belong to a family in which cancer, and especially lung cancer, is likely to occur.

If our own parents were cancer free, for example, and if their 4 parents were also cancer free (all having lived to cancer age) we probably shall have lung cancer (with cigarette irritation) only if one of our grandparents (not our parents) was a cancer hybrid in that level of our ancestry. This would give us, in other words, the right to smoke cigarettes with more or less impunity (and probable immunity) and with small danger of lung cancer.

Dr. Maude Slye, devoted more than 40 years to her study of cancer in mice. She showed that cancer could be bred into, or out of, mouse families with almost exact precision according to the rules of heredity set up by Mendel. She allowed, of course, for the factors fundamental to such research, hereditary susceptibility, either dominant or recessive and either internal or external irritation of sensitive tissues to precipitate the malignant growth. She showed not only that succeeding generations of selected cancer-susceptible mice would have growths with mendelian regularity but also that particular cancers, like lung cancer, gastro-intestinal cancer, or pelvic cancer could be "propagated" by selective breeding in the same way. And, more importantly, she demonstrated that cancer-free mice could be mated for many generations without the appearance of cancer at all.‡

From Dr. Slye's experiments in the heredity of cancer it appears that inheritance

* Lincoln, Nebraska

† Slye, Maude: Cancer and heredity *in* Studies from the Otho S. A. Sprague Memorial Institute, University of Chicago, 1910-1930

‡ Studies from the Otho S. A. Sprague Memorial Institute, Chicago, 16:63.

takes the form of susceptibility to cancer (a recessive characteristic) and not to direct inheritance of the cancer itself, (dominant). This can be demonstrated in several ways but in no better way perhaps than by the fact that Dr. Slye in her experiments has gone so far as to develop certain strains of cancer-susceptible mice in which the cancer appears not only regularly in succeeding generations but appears even in the same organs or in the same part of the body in true mendelian percentages of all the individuals studied.

In one form of cancer—cancer of the lung—which is very rare under ordinary conditions, Dr. Slye was able, by mating individuals who showed lung cancer, to develop succeeding generations in whom almost exactly correct mendelian percentages of individuals developed cancer of the lung. She has shown that this can be made to occur also for other cancer conditions by selecting parents of strains known to be susceptible to certain kinds of cancer and by selecting the offspring of these parents even though the individuals themselves do not have cancer. She was able to show succeeding generations in which each certain type and form of cancer and in certain regions of the body occurred regularly as was to be expected according to the experience in regard to other mendelian characteristics.

Dr. Slye said:

> Whenever in this laboratory two analyzed cancer free mice have been mated, it has always been possible to secure 100% cancer-free families. In such crosses no instance of cancer in the succeeding strain has ever occurred. Also, when two cancerous mice have been mated it has been possible to secure 100% cancer susceptible strains except for those mice that have died in infancy or that have been swept off by infections earlier in life than the normal age for the type of cancer to which they are predisposed. Occasionally a mouse in one of these 100% cancer strains derived from double cancerous parentage has developed a cancer when only two weeks old, although six months is an early cancer age in mice and is approximately the equivalent of about 32 years, an early cancer age in man.*

In two reports by Dr. Slye, in 1941, she deals with the type of tumor and the age at which it occurs in one series of 7,332 mice, and with 2,865 lung cancer mice out of a total of about 150,000 in the other. In all of these, with all of the points about the checking and study of every mouse, she comes up with her earlier conviction that all of these conditions are influenced by only two principal factors, that is, heredity and local tissue irritation.

The fact that individuals may have a cancer precipitated at an earlier age, or in a more aggravated form, by either internal or environmental irritations, is always recognized. But it is also pointed out that a known hybrid recessive individual may be protected, sometimes for as long as he lives, by guarding against these irritations (such as cigarettes) with sufficient care.

Dr. Slye was entirely convincing so far as her observations and findings were concerned. Her matings of the laboratory animals and her diagnoses of the growths in the generations of "cancer susceptible" families were not questioned. But, as often happens, scientists "fell out" over some of her conclusions and their own explanations as to how, or to what extent, chromosomes and genes entered into the occurrences which had taken place.

Dr. Madge T. Macklin, in a letter to me, said that Dr. Slye had thought at first that cancer susceptibility was transmitted by a single recessive gene. Many geneticists thought that she was wrong, and she abandoned that opinion. Dr. Macklin said:

> Slye was wrong in assessing all cancers to a single recessive gene (one for each type of cancer) and Little was also wrong in assuming that there was dominance. Lung cancer was thought at first to be due to a single dominant but that now is thought to be due to many genes.

* Studies from the Otho S. A. Sprague Memorial Institute, 16:963.

14

Further Studies of the Inheritance of Hand and Foot Anomalies

OLA JOHNSTON, PH.D.*

Malformations of the hands and the feet are common and of many kinds, and are clearly recognizable in individuals when present, for the abnormalities are in sharp contrast with the normal.

Snyder says that

> any finished character is the co-operative result of hereditary and environmental influences, and neither genes nor environmental influences can be solely responsible for any character.

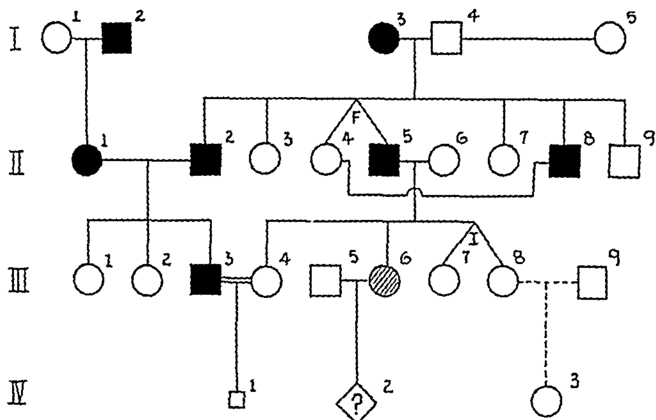
The hereditary material that a parent contributes to an offspring consists of a complement of genes. When 2 gametes that contain a set of chromosomes and genes unite, a zygote or fertilized ovum is produced. The zygote then contains 2 sets of genes, and these genes are passed by mitosis to daughter cells. Therefore, all the cells of the body will have the same genetic composition as did the zygote. The genetic constitution of man thus received determines his developmental potentialities, but a change in the germ plasm may cause an abnormal development of a character. When such a change is a permanent transmissible one, it is called a mutation. Mutations usually are harmful rather than beneficial While it is true that most genes are very stable in structure, it is probable that all congenital anomalies were produced originally by some action on the genic structures or by environmental influences or by both effects. If the anomaly is consistent, and if it is transmitted from parent to child for generations, it is believed that the anomaly is inheritable, and that it may be transmitted through dominant genes.

The pattern of development may be influenced by disease or by the environment. An example of the latter is given by Greulich and Pyle, who found that the bones of the hand and the wrist had a rather constant order in which the carpals and the epiphyses began to ossify, but that a precocious puberty, when gonadal and other hormones were present abnormally early, would accelerate the rate of ossification and skeletal development. On the other hand, decreased gonadal hormones produced by immature gonads would retard the skeletal development of the hand.

An individual may receive genes for tallness from his parents and not develop to the potentialities of his genotype when his diet is deficient in the vitamins and the minerals necessary for the formation of a normal skeleton. But no amount of adequate diet will cause him to develop into a tall person if he does not possess the genes for tallness. Here the genotype determines the height of the individual

In this study directions for the construction of a pedigree chart are given, and the results of a study of some hand and foot anomalies are presented. The malformations which are included are zygodactyly, clino-

* Denton, Texas.



= unaffected male

= unaffected female

= sex unknown

= sex unknown, number unknown

= abortive male

= parents not married

= illegitimate child

= identical twins, diagnosed

= fraternal twins, diagnosed

= twins, not diagnosed

= consanguineous marriage

= parents

= child

= male showing webbing between toes two and three

= male showing slight webbing between toes two and three

= male showing webbing between all toes of both feet

= male showing clinodactyly and streblomicrodactyly

= female showing clinodactyly and streblomicrodactyly

= male showing pes planus

= female showing pes planus

= male showing hallux valgus

= female showing hallux valgus

= male showing both pes planus and hallux valgus

= female showing both pes planus and hallux valgus

FIG. 1. *(Top)* Sample pedigree chart. Roman figures to the left indicate generations investigated. Arabic figures locate individuals. Example: III-3 = the male in the 3rd generation who married his cousin III-4. *(Bottom)* Explanation of symbols.

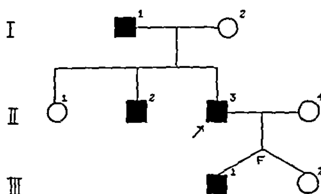


FIG. 3. Pedigree 2. A 3-generation pedigree of a family showing webbing between all toes of both feet with transmission in the male line only.

develop and elongate, the webbing decreases, until at 8 weeks only remnants remain at the base of the digits. In the case of zygodactyly the webbing is not reduced but grows outward with the toes. Stiles and Hawkins (1946) concluded that because of the embryogeny of webbed toes, zygodactyly appeared at birth and did not increase as the individual grew older.

The rarity of zygodactyly is shown by Schurmeier (1943), who reported that in the examination of 20,000 drafted servicemen, only 8 cases of zygodactyly were found, and these always between toes 2 and 3.

In the present study the pedigrees of 2 new cases of zygodactyly are presented; these were obtained through the co-operation of college students of North Texas State College. Since these families lived at a distance from Denton, Texas, it was not possible to obtain either photographs or roentgenograms of any affected individuals.

In Pedigree 1, Figure 2, 6 males show zygodactyly of toes 2 and 3. The webbing extends beyond the 2nd joint on the right foot, but not to the 2nd joint on the left foot. One male, II-5, shows only a slight webbing between toes 2 and 3. The webbing does not prevent the movement of either toe or of both toes, and it is present at birth but does not increase with the age of the person affected. Since the webbing occurred between the 2nd and the 3rd toes and no syndactylia was present in the hands, this case would be classified by Bell as Type A_1. Zygodactyly is present in the males and is transmitted from an affected male to his sons but not to his daughters; therefore, the pedigree of the family presented suggests that zygodactyly is inherited as a dominant gene, located in the y-chromosome with incomplete penetrance and variation in expression.

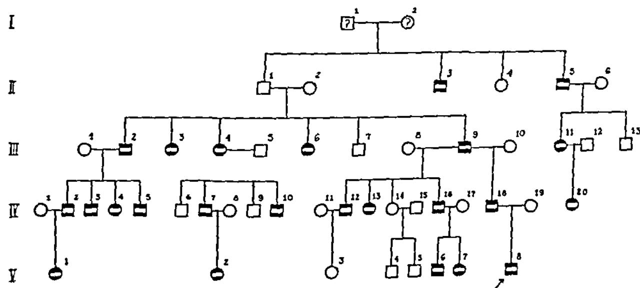


FIG. 4. A 5-generation pedigree (Pedigree 3) of a family showing clinodactyly and *streblomicrodactyly*.



FIG. 5. Hands of propositus (V-8, Pedigree 3) showing minor streblomicrodactyly.

The propositus of the 2nd family (II-3, Pedigree 2) reported as having zygodactyly gave the information that produced Pedigree 2, Figure 3. This case is of particular interest because the webbing is present between all toes of both feet and reaches almost to the tip of the toes. The skin holds the toes tightly and prevents any individual movement. The extreme zygodactyly causes the feet to be very narrow but does not impede locomotion. The anomaly is present in the males and is transmitted from affected fathers to their sons but not to their daughters. The pedigree of the family presented suggests that the zygodactylous condition of all the toes is inherited as a dominant gene located in the y-chromosome with high penetrance and no variation.

The 2 pedigrees are important, for they contribute more information on zygodactyly and give support to the dominant y-chromosome method of inheritance.

CLINODACTYLY AND STREBLOMICRODACTYLY

Stoddard (1939) clarified the nomenclature of hereditary crooked fingers by defining camptodactylia as the flexion of any finger, whereas streblomicrodactyly refers to the flexion in only the little finger. In this same article Stoddard records a family with 11 cases in 4 generations of crooked little fingers due to a short tendon. In 1 case the defect was corrected by massaging and thus stretching the tendon.

Hefner (1929) concluded that the inability to extend one or both little fingers in the region of the proximal phalangeal joint was caused by an abnormal shortening of the flexor tendon which attached to the middle phalanx of the little finger.



FIG. 6. Dorsal view of hands of propositus (V-8, Pedigree 3) showing clinodactyly.

Hersh, De Marinis and Stecher (1953),



FIG. 7. Roentgenogram of hand of propositus (V-8, Pedigree 3) showing medial view of little finger, bone structure and position in streblomicrodactyly.

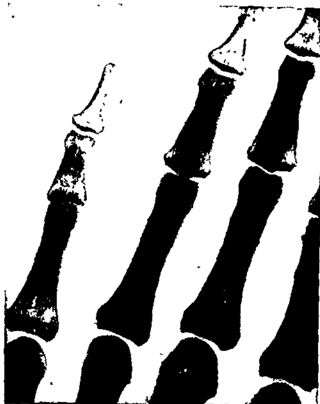


FIG. 8. Roentgenogram of left hand of propositus (V-8, Pedigree 3) showing angle of radial deflection in clinodactyly.

in a study of clinodactyly of the little finger, refer to the clinodactylous condition as a permanent radial inclination of the 5th digit caused by defective bone formation on the radial side of the middle phalanx of the 5th finger. From an analysis of their data and a review of the data of Bell (1951 and 1953), Hefner (1924, 1929, 1941), Pol (1921), Koenner (1934), Moore and Messina (1936) and Stoddard (1939) on crooked little fingers, these authors concluded that the inherited types of crooked little fingers might be divided into 3 general categories: (1) those due to incomplete ossification; (2) those due to abnormal tendons; and (3) those caused by fused ossification.

A new case of clinodactyly was made possible when the propositus (V-8) of the family pedigree (Pedigree 3) reported here, a college student, drew the author's attention to his crooked little fingers. Through his co-operation the photographs and the roentgenograms of his hands revealing the extent of the anomaly were obtained. Inquiry regarding members of the young man's family produced the pedigree. Figures 5 and 6 show the hands of the propositus (V-8, Pedigree 3). Figure 5 illustrates the condition of streblomicrodactyly, while Figure 6, a different view of the hands, shows the condition of clinodactyly. Figures 7 and 8 are roentgenograms of the propositus. Figure 7 is a medial view of the little finger in which bone structure and position in streblomicrodactyly are revealed. Figure 8 indicates the angle of radial deflection in clinodactyly of the little finger of the left hand.

Since the roentgenograms revealed little or no incomplete bone development of the radial side of the middle phalanx, clinodactyly and streblomicrodactyly are the result of a short flexor tendon attached to the middle phalanx of the little finger.

No available information could be obtained regarding crooked little fingers in I-1 and I-2 of the pedigree of the family presented here, Pedigree 3. It may be assumed that one or the other carried the gene for the anomaly. Generation II reveals 2 affected males and 1 who did not show the trait; but of his 6 children, 5 show the crooked little finger. The trait is shown in II-5, who has a daughter and a granddaughter with the anomaly. In Generation III, 6 individuals are affected, and in Generation IV 11 are affected.

Generation V reveals 5 individuals having crooked little fingers, making a total of 24 persons affected in the 5 generations.

According to Stoddard's nomenclature of hereditary crooked fingers, the anomaly in this family is streblomicrodactyly, but, according to Hersh, De Marinis and Stecher, the crooked little finger of this family belongs in their Class 2. This class includes all cases of crooked little fingers due to abnormal tendons. The pedigree (Fig. 3) reproduced here indicates that the trait is inherited as a simple mendelian dominant gene with reduced penetrance.



FIG. 9. Showing brachydactylia. (After Johnston & Davis: Am. J. Human Genet. 5:356-372)



FIG. 10. Roentgenogram showing brachydactylia. (After Johnston & Davis: Am. J. Human Genet. 5:356-372)

BRACHYDACTYLIA AND BRACHYMETAPODY

Brachydactylia is the abnormal shortness of the fingers and the toes. The condition may be caused by short phalanges or by the absence of certain phalanges in either fingers or toes, or in both fingers and toes, of an affected person. Brachymetapody is a related condition in which the phalanges are of normal length but one or more of the metacarpals or metatarsals are abnormally short.

Breitenbecker (1923) and Hefner (1924) reported cases of brachydactylia in which the terminal phalanx of the thumb of affected individuals was shortened. The trait was reported by each author to be a simple mendelian dominant.

Cutter and Nelson (1931) mentioned the occurrence of brachydactylia only in the feet of the family reported by them, and in each it appeared as a simple dominant. According to these authors, brachydactylia is one of the first mendelian traits found in man. In other families the condition may occur in the hands or the feet only, or in both hands and feet.

Brachydactylia was reported by Sayles and Jailer (1934) in 4 generations of a family in which the anomaly was regarded as a simple dominant. All affected individuals had short thumbs in which the terminal phalanx of the thumbs was shortened and widened, resulting in a squatty appearance of the whole digit.

Five generations of a large family in West Virginia with short digits were reported by Walter (1938). The trait was inherited as a simple mendelian dominant, but the expression of the gene varied from one in which a few or all fingers or toes were short to one in which both fingers and toes were short.

A case of brachydactylia in which the anomaly was transmitted as a recessive trait was reported by Cohn and Ravin (1941). The condition appeared as a bilateral symmetric shortening of the index fingers and the first toes. The hands and the feet were normal in every other respect.

McNutt (1946) suggests that many, if not all, forms of human brachydactylia are the result of the same dominant autosomal gene with reduced penetrance and varying expressivity.

Johnston and Davis (1953) reported a possible mutation in a single case of brachyphalangia in a pedigree of a family of 3



FIG. 11. Showing brachymetapody.

generations. In this pedigree the affected person has a brachydactylous condition of the hands (Fig. 9) in which all the digits are affected, except the 4th, or the ring, finger on each hand. The 1st phalanx of the right thumb, although shorter than normal, is longer than the corresponding phalanx of the left. Moreover, the thumb of the right hand is slightly crooked, whereas that of the left is relatively straight; otherwise, the 2 hands are bilaterally symmetric. Figure 10 is a roentgenogram which shows that the middle phalanx of the index, the middle and the 5th fingers of each hand is shortened considerably. The 1st and the 2nd phalanges of the index and the middle fingers on both hands are shortened, but not to the extent exhibited in the middle phalanges.

This case is included because it represents a variability of the expression of the gene for brachyphalangia. Since only one individual of this family shows brachydactyly, one cannot be certain of the cause, but, if it is a mutation, this case is evidence that mutations may occur sporadically in man.

Brachymetapody has been reported in the hands or the feet or in both hands and feet of affected individuals Gillette (1931) reported a case in which the metacarpals of the 3rd fingers of both hands were shortened. In all cases the defect was not observed



FIG. 12. Roentgenogram showing brachymetapody.

until he or she was 7 years of age. The trait was transmitted as a mendelian dominant

A case of brachymetapody, in which the 4th finger of the left hand hardly reached to the joint of the 2nd finger, was reported by Koenner (1934).

According to Stiles (1946), brachymetapody is a relatively rare anomaly. In a family of 4 generations reported by him, only the 4th metacarpal of the left hand was from birth about two thirds as long as the corresponding one of the right hand. The anomaly in this family was reported as being transmitted as an irregular dominant.

Steggerda (1942) presented an interesting case of brachymetapody in a family. The metatarsals on both feet were the only parts

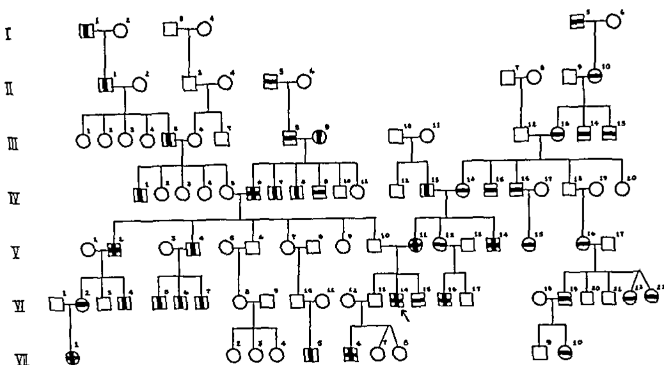


FIG. 13. A 7-generation pedigree (Pedigree 4) of a family showing pes planus and hallux valgus.

affected, the hands being normal. The trait was transmitted as a simple dominant.

Brachymetapody seems to occur more often in the metacarpals than in the metatarsals; therefore, a new case of brachymetapody is presented in which the right foot only is affected. The family whose son shows brachymetapody is a neighbor of the author. An interview with the mother and with the afflicted son produced the information. The son showing brachymetapody is the only member of a large 4-generation family that is affected. Brachymetapody is expressed by the shortening of the 4th and the 5th digits of the right foot (Fig 11). An examination of the roentgenogram (Fig. 12) reveals that metatarsals 4 and 5 are shortened, but that metatarsal 4 is much shorter than metatarsal 5, thus causing toes 4 and 5 to appear to be the same length. The abnormality does not produce any ill effects in the affected person. Since only one individual shows the trait, nothing definite can be said regarding the cause. It may represent a mutation, but if inherited the anomaly probably is the result of a single gene. The pedigree is included in this chapter as a case of a possible mutation.

HALLUX VALGUS

According to Haines and McDougall (1954), the great toe is displaced laterally in hallux valgus and usually is pronated on the head of the metatarsal; the plantar pad and the sesamoids are displaced with the digit, and the ligaments on the medial side of the joint are stretched.



FIG. 14. Showing hallux valgus in propositus (VI-14, Pedigree 4).



FIG. 15. Roentgenogram showing hallux valgus in propositus (VI-14, Pedigree 4).



FIG. 16. Showing hallux valgus in grandmother (IV-14) of propositus (VI-14, Pedigree 4).



FIG. 17. Showing hallux valgus in mother (V-11) of propositus (VI-14, Pedigree 4).



FIG. 18. Photograph showing hallux valgus in aunt (V-12) of propositus (VI-14, Pedigree 4).

Both Morgan and Riddle (1924) advanced the theory that hallux valgus might be a hereditary trait. Sorsby quotes McElvenny (1944) as concluding that hallux valgus may be hereditary and probably dominant.

Lake (1952) states that hallux valgus is produced by the effect of excessive strain thrown upon the anterior metatarsal region, and that this region is inherently weak from an evolutionary point of view and, in certain cases, unable to withstand these strains. While Lake does not indicate the probable method of inheritance for the hallux valgus, he mentions a case in which the deviation of the 1st metatarsal is an inborn characteristic, and in such cases hallux valgus is very prone to develop and then is truly congenital in origin.

The propositus of the family reported here is a graduate student in the biology department of North Texas State College. Several interviews with the young man produced the pedigree, and through his cooperation the photographs and the roentgenogram of the affected individuals were obtained.

Both hallux valgus and pes planus were present separately in members of the family of Pedigree 4 until the 2 anomalies became combined in IV-6. The anomalies continued together in V-2, V-11, V-14, VI-14, VI-16, VII-1 and VII-6. Since hallux valgus produces the greatest discomfort in the affected members of the family shown in Pedigree 4, this anomaly will be discussed. The malformation is present at birth and increases in severity with age. The condition makes it difficult to fit the individual with shoes and hampers locomotion. The photograph (Fig. 14) of the propositus (VI-14) reveals bilateral hallux valgus, and the roentgenogram (Fig. 15) of the propositus shows that the 1st metatarsal is deviated, and so is the toe, but it has not forced the 2nd toe out of its proper alignment, nor is there any overlap of the bony parts. The basal phalanx is displaced laterally, and the lateral sesamoid also is displaced laterally. Basal phalanges 2, 3 and 4 also show lateral deviation, but the terminal phalanx of the 5th *digit* deviates medially, thereby causing the digit to bend downward and to form compensatory enlargement of the 5th metatarsophalangeal joint.

Photographs of the grandmother (IV-14, Fig. 16), the mother (V-11, Fig. 17) and an aunt (V-12, Fig. 18) of the propositus show extreme lateral deviation of the digitis and well-developed bunions at the metatarsophalangeal joint.

Beginning with I-5, hallux valgus is present in each succeeding generation, and affected parents contribute the anomaly to their children, with 2 exceptions. The 1st exception is found in IV-18 and IV-19 showing no hallux valgus but having a daughter with the anomaly. This one exception may be explained by incorrect information, for IV-18 and IV-19 are deceased, and information concerning the condition of their feet depended upon recall by an elderly relative of the propositus. The 2nd exception is VI-13, who shows neither hallux valgus nor pes planus but has a son showing the combination of both.

The pedigree of the family presented suggests that hallux valgus is transmitted as an autosomal dominant with incomplete penetrance, since it is not certain that IV-18 showed the anomaly, and it is known that VI-13 does not show either anomaly.

SUMMARY

The directions for the construction of a pedigree chart are given with a sample pedigree chart illustrating the use of symbols.

Two pedigrees exhibiting zygodactyly of the feet are presented. The 1st pedigree shows the webbing present between toes 2 and 3 and extending farther on the right foot than on the left, and with the webbing on the right foot showing variations in extent. Sons only of affected fathers show the anomaly. The 2nd pedigree shows the webbing present between all toes of both feet and extending almost to the tip of the toes with no variation in expression. Affected fathers transmit the anomaly to their sons. The method of inheritance is due to a dominant gene located in the y-chromosome. In the 1st case variation of webbing is expressed, but in the 2nd case no variation is shown.

A 5-generation pedigree of a family showing slight clinodactyly and streblomicrodactyly is described, and it was found that, with one exception, all affected individuals transmitted the anomalies to their sons and daughters. Consequently, these traits are thought to be transmitted by a possible dominant gene with reduced penetrance.

Two rare anomalies are presented: (1) of a 3-generation family, in which brachydactylia is present in only 1 member (Johns-



FIG 15. Roentgenogram showing hallux valgus in propositus (VI-14, Pedigree 4).



FIG. 16. Showing hallux valgus in grandmother (IV-14) of propositus (VI-14, Pedigree 4).



FIG. 17. Showing hallux valgus in mother (V-11) of propositus (VI-14, Pedigree 4).



FIG. 18. Photograph showing hallux valgus in aunt (V-12) of propositus (VI-14, Pedigree 4).

Both Morgan and Riddle (1924) advanced the theory that hallux valgus might be a hereditary trait. Sorsby quotes McElvenny (1944) as concluding that hallux valgus may be hereditary and probably dominant.

Lake (1952) states that hallux valgus is produced by the effect of excessive strain thrown upon the anterior metatarsal region, and that this region is inherently weak from an evolutionary point of view and, in certain cases, unable to withstand these strains. While Lake does not indicate the probable method of inheritance for the hallux valgus, he mentions a case in which the deviation of the 1st metatarsal is an inborn characteristic, and in such cases hallux valgus is very prone to develop and then is truly congenital in origin.

The propositus of the family reported here is a graduate student in the biology department of North Texas State College. Several interviews with the young man produced the pedigree, and through his cooperation the photographs and the roentgenogram of the affected individuals were obtained.

Both hallux valgus and pes planus were present separately in members of the family of Pedigree 4 until the 2 anomalies became combined in IV-6. The anomalies continued together in V-2, V-11, V-14, VI-14, VI-16, VII-1 and VII-6. Since hallux valgus produces the greatest discomfort in the affected members of the family shown in Pedigree 4, this anomaly will be discussed. The malformation is present at birth and increases in severity with age. The condition makes it difficult to fit the individual with shoes and hampers locomotion. The photograph (Fig. 14) of the propositus (VI-14) reveals bilateral hallux valgus, and the roentgenogram (Fig. 15) of the propositus shows that the 1st metatarsal is deviated, and so is the toe, but it has not forced the 2nd toe out of its proper alignment, nor is there any overlap of the bony parts. The basal phalanx is displaced laterally, and the lateral sesamoid also is displaced laterally. Basal phalanges 2, 3 and 4 also show lateral deviation, but the terminal phalanx of the 5th digit deviates medially, thereby causing the digit to bend downward and to form compensatory enlargement of the 5th metatarsophalangeal joint.

Photographs of the grandmother (IV-14, Fig. 16), the mother (V-11, Fig. 17) and an aunt (V-12, Fig. 18) of the propositus show extreme lateral deviation of the digitis and well-developed bunions at the metatarsophalangeal joint.

Beginning with I-5, hallux valgus is present in each succeeding generation, and affected parents contribute the anomaly to their children, with 2 exceptions. The 1st exception is found in IV-18 and IV-19 showing no hallux valgus but having a daughter with the anomaly. This one exception may be explained by incorrect information, for IV-18 and IV-19 are deceased, and information concerning the condition of their feet depended upon recall by an elderly relative of the propositus. The 2nd exception is VI-13, who shows neither hallux valgus nor pes planus but has a son showing the combination of both.

The pedigree of the family presented suggests that hallux valgus is transmitted as an autosomal dominant with incomplete penetrance, since it is not certain that IV-18 showed the anomaly, and it is known that VI-13 does not show either anomaly.

SUMMARY

The directions for the construction of a pedigree chart are given with a sample pedigree chart illustrating the use of symbols.

Two pedigrees exhibiting zygodactyly of the feet are presented. The 1st pedigree shows the webbing present between toes 2 and 3 and extending farther on the right foot than on the left, and with the webbing on the right foot showing variations in extent. Sons only of affected fathers show the anomaly. The 2nd pedigree shows the webbing present between all toes of both feet and extending almost to the tip of the toes with no variation in expression. Affected fathers transmit the anomaly to their sons. The method of inheritance is due to a dominant gene located in the y-chromosome. In the 1st case variation of webbing is expressed, but in the 2nd case no variation is shown.

A 5-generation pedigree of a family showing slight clinodactyly and streblomicrodactyly is described, and it was found that, with one exception, all affected individuals transmitted the anomalies to their sons and daughters. Consequently, these traits are thought to be transmitted by a possible dominant gene with reduced penetrance.

Two rare anomalies are presented: (1) of a 3-generation family, in which brachydactylia is present in only 1 member (Johns-

ton and Davis); and (2) a case of brachymetapody in a family of 4 generations, in which only 1 member is affected. The 2nd case is similar to that reported by Steggerda, in that only the metatarsals are affected. This case is of particular interest, for short metatarsals occur less frequently than do short metacarpals. In both pedigrees the anomaly of each is interpreted as a possible mutation.

In the 7-generation pedigree of a family showing hallux valgus, the anomaly appears to be transmitted as an autosomal dominant with incomplete penetrance.

The cases presented of zygodactyly of the toes, clinodactyly and streblomicrodactyly of the fingers, brachydactylia of the fingers, brachymetapody and hallux valgus illustrate some of the many and varied abnormalities of the hands and the feet. The pedigrees of the families studied furnish additional evidence of the inheritance of hand and foot anomalies.

BIBLIOGRAPHY

Adams, F. H.: Hereditary deformities in man, J. Hered. 36:2-7, 1945.

Alvord, R. M.: Zygodactyly and associated variations in a Utah family, J. Hered. 38: 49-53, 1947.

Ashley, L. M.: The inheritance of streblomicrodactyly, J. Hered. 38:93-96, 1947.

Bailey, S. D. A.: A pedigree of syndactylism, J. Hered. 29:467, 1938.

Baur, E., Fischer, E., and Lenz, F.: Human Heredity, Translated by Eden and Paul, New York, Macmillan, 1931.

Bell, Julia: On hereditary digital anomalies: Part 1—On brachydactyly and symphalangism, Treasury of Human Inheritance 5:1-31, 1951.

———. On hereditary digital anomalies: Part 2—On syndactylism and its association with polydactyly, Treasury of Human Inheritance 5:33-50, 1953.

Birch-Jensen, A.: Congenital deformities of the upper extremities, Opera ex Domo Biol. Hered Human. Univ. Hafn., vol. 19, Copenhagen, Munksgaard, 1949.

Breitenbecker, J. K. Hereditary shortness of thumbs, J. Hered 14:15-21, 1923.

Castle, W. E.: Further data on webbed toes, Science 55:703-704, 1922.

Cohn, B. N. E., and Ravin, A.: An unusual case of brachydactyly, J. Hered. 33:45-48, 1941.

Cook, R.: Too few fingers—and too many, J. Hered. 26:457-462, 1935.

Cushing, H.: Hereditary anchylosis of the proximal phalangeal joints (symphalangism), Genetics 1:90-106, 1916.

Cutter, R. T., and Nelson, W. M.: A short-toed family, J. Hered. 22:163-166, 1931.

Eaton, O. N.: A summary of lethal characters in animals and man, J. Hered. 28:320-326, 1937.

Gates, R. R.: Human Genetics, New York, Macmillan, 1946.

———: Pedigrees of Negro Families, New York, Blakiston Division of McGraw-Hill, 1949.

Gillette, C. P.: An inheritable defect of the human hand, J. Hered. 22:189-190, 1931.

Glossary of Genetic Terms: J. Hered. 28:72, 1937.

Goff, C. W.: Dictionary of probable causes of congenital malformations as encountered in orthopaedic surgery *in* Am. Acad. Orthop. Surgeons Instructional Course Lectures 7: 86-97, 1950.

———: An introduction to the study of congenital malformations *in* Am. Acad. Orthop. Surgeons Instructional Course Lectures 7: 75-85, 1950.

Graham, W. C.: Congenital deformities of the hand *in* Am. Acad. Orthop. Surgeons Instructional Course Lectures 11:289-298, 1954.

Greulich, W. W., and Pyle, S. I.: Radiographic Atlas of Skeletal Development of the Hand and Wrist, p. 190, Stanford, Cal., Stanford Univ. Press, 1950.

Haines, R. W., and McDougal, A.: The anatomy of hallux valgus, J. Bone & Joint Surg. 36B:272-292, 1954.

Hall, G. A. M.: Hereditary brachydactylism and interphalangeal ankylosis, Ann. Eugenics 3:265-268, 1928.

Hefner, R. A.: Crooked little fingers, J. Hered. 32:37-38, 1941.

———: Inheritance of crooked little fingers (streblomicrodactyly), J. Hered. 20:395-398, 1929.

———: Inherited abnormalities of the fingers: I. symphalangism, J. Hered. 15:323-329, 1924.

———: Inherited abnormalities of the fingers: II. short thumbs (brachymegalodactylism), J. Hered. 15:432-439, 1924.

Hegdekatti, R. M.: Congenital malformation

of hands and feet in man, J. Hered. 30:191-196, 1939.
Hersh, A. H., De Marinis, F., and Stecher, R. M.: On the inheritance and development of clinodactyly, Am. J. Human Genet. 5: 257-267, 1953.
Hurlin, R. G.: A case of inherited syndactyly in man, J. Hered. 11:334-335, 1920.
Iltis, H.: A new case of typical brachydactyly, J. Hered. 35:145-148, 1944.
Koenner, Dora Maria: Abnormalities of the fingers, J. Hered. 25:329-334, 1934.
Lake, N. C.: The Foot, London, Baillière, Tindall & Cox, 1952.
Lewis, T.: Hereditary malformations of the hands and feet, Treasury of Human Inheritance 1:6-15, 1909.
McNutt, C. W.: Variability in the expression of the gene for brachydactyly, J. Hered. 37:359-364, 1946.
Milles, Bess Lloyd: The inheritance of human skeletal anomalies, J. Hered. 19:28-46, 1928.
Moore, W. G., and Messina, P.: Camptodactylism in its variable expression, J. Hered. 27:27-30, 1936.
Murphy, D. R.: Five successive generations of webbed-finger deformities, J.A.M.A. 84: 576-577, 1925.
Nissen, K. I.: A study in inherited brachydactyly, Ann. Eugenics 5:281-301, 1933.
Penhallow, D. P.: An unusual case of polydactylism, J.A.M.A. 91:564, 1928.
Penrose, L. S.: Inheritance of zygodactyly, J. Hered. 37:285-287, 1946.
Pipkin, A. C., and Pipkin, S. B.: Two new pedigrees of zygodactyly, J. Hered. 36:313-316, 1945.
Roberts, J. A. F.: Introduction to Medical Genetics, pp. 47-49, 197, London, Oxford, 1940.
Sayles, L. P., and Jailer, J. W.: Four generations of short thumbs, J. Hered. 25:377-378, 1934.
Schofield, R.: Inheritance of webbed toes, J. Hered. 12:400-401, 1921.
Schultz, A. H.: Zygodactyly and its inheritance, J. Hered. 13:113-117, 1922.
Schurmeier, H. L.: Congenital deformities in drafted men, Am. J. Phys. Anthropol. 5: 51-60, 1922.
Scott, W.: Syndactylism with variations, J. Hered. 24:240-243, 1933.
Snyder, L. H.: A recessive factor for polydactylism in man, J. Hered. 20:73-77, 1929.
Sorsby, A.: Clinical Genetics, pp. 268, 280, St. Louis, Mosby, 1953.
Spear, G. S.: The inheritance of flexed fingers, J. Hered. 37:189-192, 1946.
Steggerda, M.: Inheritance of short metatarsals, J. Hered. 33:233-234, 1942.
Stiles, K. A., and Hawkins, D. A.: An inheritance of zygodactyly, J. Hered. 37:16-18, 1946.
Stoddard, S. E.: Nomenclature of hereditary crooked fingers, J. Hered. 30:511-512, 1939.
Strandskov, H. H.: An outline of the principles of human genetics in *Am. Acad. Orthop. Surgeons Instructional Course Lectures 7:63-68, 1950.*
Straus, W. L.: The nature and inheritance of webbed toes in man, J. Morphol. 41:427-439, 1926.
Walter, Mable R.: Five generations of short digits, J. Hered. 29:143-144, 1938.

Studios Additional Super le Hereditate de Anomalias de Mano e Pede

Summario in Interlingua

Ex le longe e variate lista de anormalitates de mano e pede, casos representative es describite de zygodactylia del digitos del pede, clinodactylia e streblomicrodactylia del digitos del mano, brachydactylia del digitos del mano, brachymetapodia, e halluce valge. Inter le duo arbores genealogic con zygodactylia del pede, le un monstra palmation inter le secunde e tertie digito del pedes. Le palmation es plus extense al pede dextere que al pede sinistre; al pede dextere illo es characterisate per variationes del extension. Le secunde arbore genealogic monstra plamation inter omne le digitos de ambe pedes. Le palmation se extende quasi usque al puncta del digitos e non varia in su extension. In ambe casos, patres zygodactylic transmitte le condition a lor filios. Ergo le mechanismo de hereditage involve un gen dominante in le chromosoma y.

Es describite un arbore genealogic de cinque generationes con leve grados de clinodactylia e streblomicrodactylia. Con un exception, omne le individuos afficite trans-

mitteva le anomalia a lor filios e filias. Ergo il pare que iste tractos es transmittite per un possibile gen dominante con reducite penetrantia.

Es presentate duo rar anomalias. Le prime es un familia de tres generationes con brachydactylia in solmente un membro (Johnston e Davis). Le secunde es un familia de quatro generationes in que un sol membro exhibi brachymetapodia, e solmente le ossos metatarsal es afficite. Le secunde de iste casos es de interesse special, proque curte metatarsales occurre minus frequentemente que curte metacarpales. In ambe iste arbores genealogic le anomalia es interpretate como un possibile mutation.

In le arbore genealogic de septe generationes con halluce valge, il pare que le anomalia es transmittite como dominante autosomal con penetration incomplete.

Le anomalias del arbores genealogic hic studiate adde nove datos a nostre cognoscentias del hereditage de anomalias de mano e pede.

Section II

GENERAL ORTHOPAEDICS

of the same age, thereby rendering themselves vulnerable to harmful stresses that produce joint hemorrhages.

The patient's personality and emotional status play a significant role in the incidence of joint hemorrhages. Those who are placid and dull suffer fewer hemorrhages than those who are active, apprehensive and emotionally unstable.

The joints implicated most often in the order of frequency are knee, elbow, ankle and hip. This study reveals that specific factors play a part in the selectivity. In addition to the severity of the blood defect, these factors comprise (1) vulnerability of the joint, (2) anatomic peculiarities and (3) occupation and other activity demanded of the joint incident to function. The joints especially vulnerable to injury are the knees, the elbow and the ankle. This is borne out clinically by the high incidence of hemarthroses encountered in these joints. Some joints possess anatomic peculiarities that predispose them to intra-articular bleeding. This is particularly true of joints whose stability depends upon soft tissue structures such as ligaments and tendons rather than on their bony configuration. The knee joint exemplifies this last observation. Its anatomic construction makes it totally dependent upon the integrity of the surrounding ligaments, capsule and tendons for its stability. It is interesting to note that once the forementioned factors initiate hemarthrosis in a particular joint, that joint usually suffers repeated hemorrhages. It appears that one hemarthrosis predisposes the affected joint to another. This is true regardless of the severity of the blood defect. It is not uncommon to observe one knee or one shoulder joint severely impaired because of repeated hemarthroses, while t o posite knee or shoulder exhibits no and has been the site of very fe or- rhages.

FACTORS RESPONSI DEFO ES OF J

In lite s consta ment that in spite of repeated and severe hemorrhages in joints, little functional disability occurs. This is an erroneous observation. Critical evaluation of such joints in all instances reveals varying degrees of loss of the normal arcs of joint motion and varying grades of deformities. However, it is true that in most instances the loss in motion is a gradual process, and the patient makes the necessary adjustments to the reduced arcs of motion so that he is not handicapped seriously. In some instances the dysfunction of the affected joint is so great that it is impossible to make the necessary adjustment; hence, severe disability ensues.

The factors responsible for flexion deformities following acute massive hemorrhages are (1) pain and (2) muscle spasm. The pain results from acute distention of the capsular and the pericapsular tissues and from the irritating effect of the blood elements on the subsynovial tissues. During this acute phase the joints are distended markedly and lose their normal configuration; they exhibit increased local temperature and are painful on palpation. The joints soon exhibit on palpation a characteristic firm rubbery consistency resulting from thickening of the synovial, the capsular and the pericapsular tissues and infiltration of these tissues with blood. These features are readily discernible in the more superficial joints, as the knee, the elbow and the ankle. Occasionally in the shoulder the hemorrhage may be so extensive as to dislocate the head of the humerus.

With cessation of bleeding, the intra-articular blood is absorbed completely. However, repeated insults of this nature produce permanent alteration in the soft tissue elements of the 'oint and in those about the joint; also itant changes occur in the cartil d the osseous elements, which, to the soft tissue abnormalities, d deformities. The essential c soft tissue comprise depositio ood el in the synovial ynovial nd in- filtration and ular

tissues by blood. This process is followed by a cellular response resulting in increased vascularity, hyperplasia of the synovial membrane and thickening of the capsular and the pericapsular tissues. The process is not unlike a reparative process producing marked and diffuse fibrous tissue proliferation, which on maturity leads to the formation of dense scar tissue. Then the soft tissue elements lose their elasticity and become scarred and contracted.

Although massive hemorrhages into joints are responsible for many of the unfavorable sequelae noted above, even more important are repeated small hemorrhages that cause only minimal distention of the soft tissues and produce symptoms that are tolerated by the patient without his seeking medical aid. The pain is not of sufficient severity to limit the usual activities of the patient; and although some restriction of motion occurs, and although the joint is held in slight flexion and guarded by the patient, the dysfunction is tolerated well. These small hemorrhages are more frequent by far than massive hemorrhages, and they are a constant source of irritation to the synovial membrane and the articular cartilage. It is reasonable to conclude that they play a more important role in the formation of permanent changes than single massive hemorrhages occurring at long intervals. Also, they differ in their clinical manifestations from massive intra-articular bleeding in other ways; they produce no local increase in temperature and no constitutional reaction such as an elevation in the body temperature or a leukocytosis. These features are encountered not infrequently in massive hemarthroses.

Joint deformities may be secondary to massive hemorrhages in muscles and in fascial planes. Although not common, the authors have observed several severe hip-flexion deformities following massive and repeated hemorrhages into the psoas muscle. One patient developed a severe claw-hand deformity following hemorrhage in the palm of the hand; another exhibited a fixed adduction deformity of the thumb following



Fig. 1. Note the fixed adduction deformity of the thumb following hemorrhage into the hyperthenar eminence and the palm of the hand. Also note the fixed flexion deformity of the 3rd finger.

hemorrhage into the hyperthenar eminence (Fig. 1); and still another developed cavus deformity of both feet following hemorrhages into the fascial planes of the plantar aspects of both feet. In the literature a case of Volkmann's contracture was reported by Thomas following a massive hemorrhage that included the entire arm.

In rare instances deformities may be the result of massive joint or soft tissue hemorrhages that result in necrosis of the affected tissues. Also, the great tension in the part produced by the bleeding may result in obliteration of the main arterial trunks of the limb distal to the site of bleeding producing varying degrees of gangrene of the distal part. Thomas reports a case of gangrene of the foot following an extensive hemor-

of the same age, thereby rendering themselves vulnerable to harmful stresses that produce joint hemorrhages.

The patient's personality and emotional status play a significant role in the incidence of joint hemorrhages. Those who are placid and dull suffer fewer hemorrhages than those who are active, apprehensive and emotionally unstable.

The joints implicated most often in the order of frequency are knee, elbow, ankle and hip. This study reveals that specific factors play a part in the selectivity. In addition to the severity of the blood defect, these factors comprise (1) vulnerability of the joint, (2) anatomic peculiarities and (3) occupation and other activity demanded of the joint incident to function. The joints especially vulnerable to injury are the knees, the elbow and the ankle. This is borne out clinically by the high incidence of hemarthroses encountered in these joints. Some joints possess anatomic peculiarities that predispose them to intra-articular bleeding. This is particularly true of joints whose stability depends upon soft tissue structures such as ligaments and tendons rather than on their bony configuration. The knee joint exemplifies this last observation. Its anatomic construction makes it totally dependent upon the integrity of the surrounding ligaments, capsule and tendons for its stability. It is interesting to note that once the forementioned factors initiate hemarthrosis in a particular joint, that joint usually suffers repeated hemorrhages. It appears that one hemarthrosis predisposes the affected joint to another. This is true regardless of the severity of the blood defect. It is not uncommon to observe one knee or one shoulder joint severely impaired because of repeated hemarthroses, while the opposite knee or shoulder exhibits no alterations and has been the site of very few or no hemorrhages.

FACTORS RESPONSIBLE FOR DEFORMITIES OF JOINTS

In literature one finds constantly the statement that in spite of repeated and severe hemorrhages in joints, little functional disability occurs. This is an erroneous observation. Critical evaluation of such joints in all instances reveals varying degrees of loss of the normal arcs of joint motion and varying grades of deformities. However, it is true that in most instances the loss in motion is a gradual process, and the patient makes the necessary adjustments to the reduced arcs of motion so that he is not handicapped seriously. In some instances the dysfunction of the affected joint is so great that it is impossible to make the necessary adjustment; hence, severe disability ensues.

The factors responsible for flexion deformities following acute massive hemorrhages are (1) pain and (2) muscle spasm. The pain results from acute distention of the capsular and the pericapsular tissues and from the irritating effect of the blood elements on the subsynovial tissues. During this acute phase the joints are distended markedly and lose their normal configuration; they exhibit increased local temperature and are painful on palpation. The joints soon exhibit on palpation a characteristic firm rubbery consistency resulting from thickening of the synovial, the capsular and the pericapsular tissues and infiltration of these tissues with blood. These features are readily discernible in the more superficial joints, as the knee, the elbow and the ankle. Occasionally in the shoulder the hemorrhage may be so extensive as to dislocate the head of the humerus.

With cessation of bleeding, the intra-articular blood is absorbed completely. However, repeated insults of this nature produce permanent alteration in the soft tissue elements of the joint and in those about the joint; also, concomitant changes occur in the cartilaginous and the osseous elements, which, together with the soft tissue abnormalities, produce fixed deformities. The essential changes in the soft tissue comprise deposition of the blood elements in the synovial and the subsynovial tissues and infiltration of the capsular and the pericapsular

tissues by blood. This process is followed by a cellular response resulting in increased vascularity, hyperplasia of the synovial membrane and thickening of the capsular and the pericapsular tissues. The process is not unlike a reparative process producing marked and diffuse fibrous tissue proliferation, which on maturity leads to the formation of dense scar tissue. Then the soft tissue elements lose their elasticity and become scarred and contracted.

Although massive hemorrhages into joints are responsible for many of the unfavorable sequelae noted above, even more important are repeated small hemorrhages that cause only minimal distention of the soft tissues and produce symptoms that are tolerated by the patient without his seeking medical aid. The pain is not of sufficient severity to limit the usual activities of the patient; and although some restriction of motion occurs, and although the joint is held in slight flexion and guarded by the patient, the dysfunction is tolerated well. These small hemorrhages are more frequent by far than massive hemorrhages, and they are a constant source of irritation to the synovial membrane and the articular cartilage. It is reasonable to conclude that they play a more important role in the formation of permanent changes than single massive hemorrhages occurring at long intervals. Also, they differ in their clinical manifestations from massive intra-articular bleeding in other ways; they produce no local increase in temperature and no constitutional reaction such as an elevation in the body temperature or a leukocytosis. These features are encountered not infrequently in massive hemarthroses.

Joint deformities may be secondary to massive hemorrhages in muscles and in fascial planes. Although not common, the authors have observed several severe hip-flexion deformities following massive and repeated hemorrhages into the psoas muscle. One patient developed a severe claw-hand deformity following hemorrhage in the palm of the hand; another exhibited a fixed adduction deformity of the thumb following hemorrhage into the hyperthenar eminence (Fig. 1); and still another developed cavus deformity of both feet following hemorrhages into the fascial planes of the plantar aspects of both feet. In the literature a case of Volkmann's contracture was reported by Thomas following a massive hemorrhage that included the entire arm.



FIG. 1. Note the fixed adduction deformity of the thumb following hemorrhage into the hyperthenar eminence and the palm of the hand. Also note the fixed flexion deformity of the 3rd finger.

In rare instances deformities may be the result of massive joint or soft tissue hemorrhages that result in necrosis of the affected tissues. Also, the great tension in the part produced by the bleeding may result in obliteration of the main arterial trunks of the limb distal to the site of bleeding producing varying degrees of gangrene of the distal part. Thomas reports a case of gangrene of the foot following an extensive hemor-



Fig. 2. Observe the massive necrosis of tissues of the anterior aspect of the knee joint following an extensive hemorrhage into the joint In spite of repeated aspirations and compression, the severe tension in the joint was not reduced sufficiently to prevent tissue necrosis (Tocantins, L. M.: Progress in Hematology, vol 1, New York, Grune & Stratton)

rhage into the entire leg. The authors have observed an extensive hemorrhage into the knee joint that ruptured through the joint capsule and implicated the entire quadriceps apparatus; this was followed by massive necrosis of the tissues. In a heroic attempt to save the limb, a thorough débridement of the area was performed, and the defect was covered with split thickness skin grafts taken from the patient. The patient made an uneventful recovery. (Figs. 2 & 3.)

Muscle imbalance is another factor that may be responsible for development of fixed flexion deformities. This is especially true of the knee. We are all cognizant of the problems posed by the quadriceps muscle when the knee is injured. The resulting atrophy often is a major deterrent to rapid restoration of normal function. In the hemophiliac, repeated hemarthroses of the knee joint are associated invariably with pronounced atrophy and loss of power in the quadriceps muscle; hence, this muscle is unable to cope with the strong, and often shortened, hamstring muscles. In face of such a situation it becomes obvious that a flexion deformity of the joint is inevitable.

The first alterations in the articular cartilage comprise thinning and erosion of the superficial layers. Later, these areas are invaded by granulation tissue from the marrow spaces of the cancellous bone. At the



Fig. 3. This is the same joint as that depicted in Figure 2. Note that following excision of the necrotic soft tissue, a split-thickness graft was applied covering the entire denuded area. (Tocantins, L. M.: Progress in Hematology, vol. 1, New York, Grune & Stratton)



Fig. 4. Note the punched-out areas in the cancellous bone and in the subchondral areas of both the tibia and the femur. These cystlike areas may attain giant proportions.

peripheries the cartilage also is invaded and overlapped by a panus of hyperplastic vascular synovial and subsynovial tissues. Subchondral cysts and cysts form at a distance from the surface; atrophy of the subchondral bone is severe and leads to fracture compression and flattening of the articular surfaces. In some cases excrescencies comprising newly formed cartilage and immature bone are produced at the margins of the joints. Furthermore, repeated hemarthroses may cause an irregular increase in the width of the epiphysis, producing characteristic knobbiness of the joints. All the abovementioned alterations result in marked incongruity of the articular surfaces, restriction of motion and, eventually, fixed deformities. These observations have been recorded by Key and confirmed by us in joints that were explored surgically and in joints that were obtained post mortem. Also, these changes have been reproduced in animal experiments. The experimental study will be presented in a subsequent report.

ROENTGENOLOGIC FEATURES

The roentgenologic features of this disorder have been recorded by many observers. Notably among these are Neumann, Montonari, Engels, Doub and Davidson, Key, Klason, Rypins and Johnson *et al.* Although the findings are not pathopneumonic of the disease, there are certain features which, when noted in the male sex, should permit a presumptive diagnosis of hemophilic involvement. The roentgenologic findings do not parallel the clinical findings, nor do they indicate the degree of dysfunction except in the cases exhibiting far-advanced degenerative changes. This is true because alterations in the soft tissue elements of the joint may be playing the major role in producing the deformity and the disability. It is not uncommon to encounter joints with fixed deformities that exhibit minimal or no osseous or cartilaginous abnormalities in the roentgenograms.

Except in rare instances the changes observed in joints are governed by the number of previous massive hemorrhages and subclinical small hemorrhages; this is true regardless of the age of the patient. Of course, the older the patient becomes, the more readily will advanced changes be discernible roentgenologically, provided that he has had repeated and frequent joint hemorrhages. The most significant findings are defects often described as cysts in the subchondral and in the cancellous portions of the affected bone and increased density in the synovial and the capsular tissues. Some of the defects appear to be continuous with the joint cavity, while others are at a distance from the articular surface. (Fig. 4.) How-



Fig. 5. The joint capsule of both hip joints has undergone complete calcification and ossification.



Fig. 6. Note that the bone ends comprising the joint are broader than normal and that the joint space exhibits varying degrees of narrowing. Also note the giant-sized cysts in the medial condyle of the tibia.

ever, not all hemophilic joints disclose these characteristic punched-out depressions in the articular surface and the cystlike areas in the subchondral and the cancellous bone. They vary in size and may approach giant proportions. The shadows in the soft tissues vary in density; in some instances they approach the density of calcification of the tissues. They depict sharply the outline of the affected joint. In one case the joint capsule of both hips revealed marked calcification and stood out in sharp contrast with the surrounding tissues. (Fig. 5.) In most instances the increase in density of the synovial and the capsular tissues is the result of the diffuse hyperplastic proliferative process that exists in the tissues. In superficial joints such as the knees, the wrists and the elbows, this process is readily demonstrable on physical examination of the part. In addition to the proliferative changes, the increase in density is the result of disposition of iron pigment in the synovial and the subsynovial tissues. This is particularly true in joints which have been the site of repeated hemorrhages. In acute hemorrhage in joints which are implicated for the first time or have been the site of infrequent hemorrhages, the joint cavity is distended markedly and is demonstrated readily in the roentgenograms; it casts a uniform haziness demarcated clearly from the surrounding tissues. The shadow is produced by the blood in the joint cavity and in the synovial and the subsynovial tissues. Occasionally massive hemorrhages may produce a subluxation or even dislocation of the joint. This phenomenon was observed by the authors only in the shoulder joint.

In most instances following repeated hemorrhages, permanent alterations which in a



FIG. 7. Observe the angular deformity of the knee joint resulting from collapse of a giant cyst.

measure simulate osteoarthritis occur in the cartilaginous and the osseous elements of the joint. The bony ends comprising the joint become broader than normal, and the joint space exhibits varying degrees of narrowing, or it may be obliterated. (Fig. 6.) Only rarely is bony ankylosis encountered. Bony ankylosis was noted in only 2 cases, and the subtalar joint was implicated in both. The articular surfaces are irregular and may exhibit numerous punched-out defects not unlike those encountered in gout. As remarked previously, cystlike areas varying in size also are noted in the subchondral and in the cancellous bone. Collapse of the intervening bony trabeculae may produce angular deformities of the joint; this feature is noted not infrequently in the knee joint. (Fig. 7.) In advanced cases sharply delineated marginal spurs and osteophytes are found in close proximity to the articular surface. Spurring of the tips of the olecranon, the coronoid process of the ulna, the poles of the patella and the tibial spines is a common finding. A feature peculiar to the knee joint is a broadening and a deepening of the intercondylar notch. During the active stage of the arthritis, particularly in young patients, varying degrees of atrophy of the bone ends are discernible. Usually this is associated with coarsening of the bony trabeculae, which in some instances stand out in bold relief. In older patients in whom the disorder is stabilized or quiescent, the bony atrophy disappears and the trabeculae are less coarse; then the bony architecture tends to approach normalcy.

Frequently in the advanced stages of arthritis, in addition to the alterations recorded above, the joints reveal pronounced



FIG. 8 Observe the pronounced subchondral sclerosis that is found in the subchondral area of both the femur and the tibia.

subchondral sclerosis. (Fig. 8.) Invariably this is associated with marked marginal spurring, irregular craterlike defects in the articular surface and cysts of varying size in the subchondral and the cancellous regions of the bones. Occasionally irregular areas of increased density, spotty in distribution and following no particular pattern, are found in the shafts of the long bones at a distance from the joint. These may be calcified intra-osseous hemorrhages. It is interesting to note that cystic abnormalities may be the outstanding roentgenographic features in some cases and insignificant or absent in others. However, if the changes are noted in one joint, invariably they will be present in other joints. In some instances subchondral sclerosis is the pertinent bone change; this usually is encountered in joints affected severely by the arthropathy.

Irregular subperiosteal ossification following subperiosteal hemorrhages are encountered occasionally. In this study the fibula disclosed this abnormality most frequently.

The effect of repeated hemarthroses on growing epiphyses raises some inquiries of special clinical significance. What alterations occur in an epiphysis and diaphysis following repeated joint hemorrhages over periods of many years? Caffey and Schlesinger are of the opinion that acceleration of development and overgrowth occur. In this study several alterations were noted. In some instances rapid increase in size of the osteogenic growth center is readily discernible. This is noted frequently in the epiphyses comprising the elbow joint, particularly the head of the radius. (Fig. 9.) In most instances irregular overgrowth of the epiphyses in all their dimensions is a common finding. This is responsible for the characteristic knobbiness these joints exhibit, and it is most readily discernible in the knee joints. On the other hand, premature closure may occur of a portion of the epiphyseal plate or all of it, resulting in a shortening of the bone or angular deformities. No case in this series revealed discrepancy in bone lengths



Fig. 9. Observe the increase in size of the epiphysis of the head of the radius on the left as compared with that of the right joint.

that could be attributed to overstimulation at the epiphyseal plate. The presence of subchondral defects and flattening of the pressure epiphyses such as the capital epiphysis of the femur and the talus produces roentgenographic features that resemble closely those of the osteochondroses.

PATHOLOGY

The gross and the microscopic appearances of the tissues in hemophilic arthropathy have not been recorded frequently in the literature. Key described the synovial tissues obtained from a knee joint that was operated upon because of an erroneous diagnosis. Freund and Reinecke and Wohlwill described the microscopic changes in cartilage and bone, and König recorded the gross alterations in the hemophilic joints. We have been fortunate in obtaining several joints (2 knee joints and 2 humeral heads with the musculotendinous cuff attached) from a hemophiliac 27 years of age who came to autopsy. In addition, as will be discussed



FIG. 10. This is the right humeral head of a hemophiliac, 27 years of age. Note that the articular surface is entirely denuded of cartilage; the subchondral bone is exposed and pitted, and shows extensive spur formation. Also, observe the advanced changes that have occurred on the inner aspect of the musculotendinous cuff. The cuff is laminated and in some areas is covered by villus formations arising in the synovial membrane.

more fully subsequently, specimens of joint tissues were obtained from a hemophiliac on whom an arthrodesis of the knee joint was performed. This material forms the basis of this report.

MACROSCOPIC FINDINGS

HUMERAL SIDE OF THE RIGHT SHOULDER JOINT (FIG. 10)

The size of the articular surface of the head of the humerus is reduced by one third of its normal size. The entire surface is stained a chocolate color, and its periphery is irregular in outline and has receded from the soft tissues. Along the medial and the inferior aspects of the humeral head, the periphery presents a sharp, serrated, bony osteophyte which is free of soft tissue attachment and is continuous with the subchondral bone. No vestige of hyaline cartilage remains. The subchondral bone is eburnated and peppered with numerous craterlike defects that vary in size and in shape. Some areas in the upper one half of the articular surface reveal complete loss of subchondral bone; here the cancellous bone is exposed, and it exhibits also irregular punched-out defects that communicated with the joint cavity. Many of the defects in the cancellous bone exhibit a smooth, firm, compact inner surface, leading one to conclude that the contents of the cyst were under great pressure, thereby producing collapse and condensation of the cancellous trabeculae forming its walls. Several of the defects in the cancellous bone appear to undermine the overlying subchondral bone. It was interesting to note that this specimen failed to show macroscopically any fibrous tissue proliferation growing from the defects in the subchondral and the cancellous bone. This is in marked contrast with the alterations noted in the knee joints of this same individual, *which exhibited pronounced fibrous tissue* proliferation in some areas of the articular surfaces of the femoral condyles and the patella. The reduced size of the humeral head appears to be the result of gradual disintegration and erosion of its cartilaginous and osseous elements, more pronounced along its superior and posterior borders.

The soft tissue elements vary in color from reddish-brown to chocolate. The stained synovial membrane is thin and exhibits fraying and some synovial tabs. Minimal villus formation is noted in the anterior aspect of the joint. The villi are long and slender with fine pedicles. The glenohumeral ligaments are thicker and broader than those in the left shoulder, the subscapularis recesses are obliterated because the ligaments are adherent to the capsule, and the openings of the recesses are reduced in size by proliferation of the fibrous tissue in the subsynovial layer. In the superior aspect of the joint, in the region of the supraspinatus and the infraspinatus tendons, the synovial membrane is absent. Here the thickened, frayed and

discolored tendon fibers protrude through the defect in the synovial membrane. The entire musculotendinous cuff shows advanced degenerative changes comprising shredding, lamination and fibrous tissue proliferation. It is interesting to note that the cuff has receded from 1 to 1½ cm. from the periphery of the articular surface of the humeral head. The intra-articular portion of the biceps tendon is absent in this specimen. From the fact of the marked changes noted, it is reasonable to assume that the tendon degenerated completely and was absorbed.

Humeral Side of the Left Shoulder Joint (Fig. 11)

In contrast with the profound alteration observed in the right shoulder, gross examination of the left shoulder fails to show any abnormalities. The head of the humerus is of normal shape and size, and its articular surface is covered by normal hyaline cartilage. The synovial membrane is smooth, thin, glistening and not stained by blood pigments. The glenohumeral ligaments are well delineated and covered by normal synovial membrane. No tabs, shredding or villus formation of the synovialis is demonstrable. The musculotendinous cuff is in close proximity to the periphery of the articular surface of the humeral head and exhibits no gross degenerative changes. The biceps tendon is of normal appearance and occupies its anatomic position in the intertubercular groove.

It is difficult to conceive that in a hemophiliac one shoulder joint could exhibit such profound changes and the other shoulder joint show no alterations. This observation emphasizes the role that trauma incident to function must play in initiating hemorrhage in the joints of hemophiliacs. In hemophiliacs it is not uncommon to observe changes of greater severity in the joints of the dominant extremity.

Right Knee Joint (Fig. 12)

The entire synovial lining and the articular surfaces of all the bony joint elements are varying shades of chocolate color; the cruciate ligaments and the infrapatellar fat pads are stained deeper shades than the surrounding tissues. Section of the anterior horns of both menisci and of the quadriceps tendon reveals that the entire thickness of these structures is permeated by blood pigments. The articular cartilage of the femoral condyles, the tibial plateaus and the patella reveals the same degree of involvement. The cartilage is edematous, wavy and shredded. Numerous fine villi cover the surface; these are more numerous on the surface of the femoral condyles and the tibial plateaus than on the surface of the patella. The cartilage is pitted severely and also exhibits clefts and tissues of varying size and depth from which villi project. It is interesting to note that no areas of erosion to the level of the subchondral bone are noted; also, no areas of eburnation are discernible such as are noted in the right shoulder of this person.



Fig. 11. This is the left humeral head of the same person depicted in Figure 10. Note that the humeral head and the inner aspect of the musculotendinous cuff appear to be grossly normal.

The peripheries of the femoral condyles disclose large well-developed bony osteo-



FIG. 12. A knee joint of a hemophiliac, 27 years of age. Note the extensive alterations that have occurred in the femoral condyles and in the articular surfaces of the tibial condyles. Many areas of the articular surfaces are denuded completely of cartilage. Fibrous tissue proliferation is pronounced, and numerous fine villi cover the surfaces of both bones. The cartilage is pitted and exhibits many clefts and fissures of varying size. The synovial lining is fibrotic and covered by lumps of villus formations. The peripheries of the femoral condyles disclose large well-developed bony osteophytes. These alterations also are noted at the margins of the patella. In this instance the suprapatellar pouch was obliterated completely by numerous fine villi varying in length and covering the entire synovial lining. The suprapatellar and the infrapatellar fat pads, the menisci and the cruciate ligaments also are covered by numerous villi and show evidence of marked fibrous tissue proliferation.

phytes covered with cartilage. Similar changes are noted around the margin of the patella. However, in the patella the excrescencies exhibit a more irregular pattern.

Soft tissue alterations in this joint are pronounced. The synovial membrane and the subsynovial tissue are thickened markedly Numerous fine villi, varying in length, cover the entire synovial lining and practically obliterate the suprapatellar pouch. The menisci and the cruciate ligaments are stained with blood pigments, and the intercondylar notch is obliterated by thickened hyperplastic synovial tissue. The superior and the infrapatellar fat pads are thickened, matted and covered with numerous villi of varying sizes and shapes.

Although this joint presents many features which are similar to those encountered in osteoarthritis, there are other features which are peculiar to hemophilic arthropathy. The changes in the synovialis, the capsule and the pericapsular tissues of a hemophilic joint occur early, whereas in osteoarthritis they occur later in the disorder. Moreover, the intensity of the response of these tissues in hemophilic joints surpasses by far those observed in osteoarthritis. In the latter, the hyperplastic abnormalities are confined to the synovialis, the intracapsular ligaments and the capsule; in the former, these changes also implicate the pericapsular tissues such as fasciae, ligaments, muscles and tendons Another point of difference between the two arthropathies is the uniform and the widespread involvement of the articular cartilage in hemophilic joints. It appears that the entire articular surface is exposed to the same noxious agent, and the subsequent degen-



FIG. 13. Observe that in some areas the entire thickness of the cartilage has disintegrated completely and that the surface is in close proximity to the subchondral bone and the marrow spaces.



FIG. 14. Note that Weichselbaum's lacunae are distinctly discernible. There is considerable fragmentation of the cartilage overlying the lacunae. In some areas the lacunae communicate directly with the joint cavity, giving the articular surface a serrated and pitted appearance.

erative process progresses uniformly in all regions of the articular cartilage. This is not true in osteoarthritis, in which the changes in the articular cartilage are more or less spotty in nature; first one area is implicated in the process, then another. Only in the very advanced stages of the disease does one observe widespread involvement of the articular surfaces, and even in this phase some areas are affected less than others.

LEFT KNEE JOINT

Essentially the alterations noted in this joint are similar to those observed in the right knee. The anterior compartments of the femorotibial joint were occluded partially by overgrowth of fibrous tissue in the infrapatellar region and in the intercondylar notch It was interesting to note that the outer tibial plateau had collapsed and that an angular deformity of the joint had occurred so that the outer tibial femoral condyle was on a lower plane than the inner condyle. Collapse of the outer tibial condyle is depicted more clearly in a coronal section through the joint. The sections of the specimen bring into view the large defect immediately below the articular surface of the tibial plateau which permitted the collapse. In addition, the posterior compartment is visualized; it is obliterated by dense fibrous tissue. Defects, varying in size, are observed in the cancellous portion of both femoral condyles; undoubtedly these are portrayed roentgenographically as cysts.

MICROSCOPIC FEATURES

ARTICULAR CARTILAGE

The outstanding features are the extensive degenerative changes which have occurred in the hyaline cartilage. This structure exhibits marked fibrillation of its collagen fibers, fragmentation and thinning. In some areas the entire thickness of the car-



FIG. 15. Note in this specimen that the entire thickness of the cartilage is replaced by vascular proliferating granulation tissue that extends from narrow spaces.



FIG. 16. In this specimen many marrow spaces adjacent to the subchondral bone contain an abundance of vascular proliferating connective tissue that seems to cause resorption of the surrounding bone trabeculae.

tilage exhibits complete disintegration. (Fig. 13.) In other areas the presence of Weichselbaum's lacunae and the fragmentation of the cartilage overlying the lacunae are clearly discernible. Now the lacunae communicate with the joint cavity and give the articular surface a serrated or pitted appearance. (Fig. 14.) Similar changes are noted in the deep layers of the cartilage. Here large cell nests or lacunae communicate eventually with the marrow spaces of the cancellous bone. In other areas the cellular elements have disappeared almost completely, while the alterations in the hyaline matrix result in separation of the collagenous bundles. The deep bundles now are seen running in a direction radial to the articular surface, whereas the superficial bundles are parallel to the surface.

At the margins of the articular cartilage, the hyperplastic synovial membrane extends for varying distances and dips into the degenerated cartilage, converting it into an area of vascular connective tissue throughout which remnants of cartilage tissue are dispersed.

It is observed that several areas of disintegration of the entire thickness of the cartilage are replaced by vascular proliferating granulation tissue extending from the marrow spaces. (Fig. 15.) In this hyperplastic tissue the fragments of the hyaline cartilage still are discernible. Such areas may well depict one mode of formation of subchondral cysts so characteristic of hemophilic arthritis. It is reasonable to assume that with complete absorption of the remaining fragments of the articular cartilage, a defect remains in the bone end devoid of osseous or cartilaginous elements but containing granulation tissue; this defect now communicates with the joint cavity. Con-

tinued activity of this tissue causes further absorption of the surrounding cartilage and bone, thereby increasing the size of the defect. On the other hand, the granulation tissue may undergo maturation, forming fine strands of fibrous tissue that project into the joint cavity. Such changes were noted in the macroscopic study of the hemophilic joints described previously.

Subchondral and Cancellous Bone

Marked irregular thinning of the subchondral bone is noted; in some areas the calcified layer of cartilage rests on the marrow spaces of the cancellous bone. Even more pronounced are the atrophy and the fragmentation of the bony trabeculae of the cancellous bone. This permits coalescence of the small marrow spaces and formation of larger spaces. It is interesting to note that many marrow spaces adjacent to the subchondral bone contain an abundance of vascular proliferating connective tissue which appears to cause absorption of the surrounding bony trabeculae. (Fig. 16.) This process may be responsible for the formation of cysts below the level of the articular cartilage. As noted previously, the process also may implicate the overlying cartilage; with disintegration and absorption of the cartilage the cyst gains communication with the joint cavity. There is no evidence of recent intracancellous hemorrhage that might be interpreted as the initiating agent of this process. In this instance it appears that the degenerating cartilage may be the exciting factor. However, one must admit that hemorrhage within the cancellous bone may occur and initiate a similar pathologic process that results in destruction of atrophic bony trabeculae and the formation of intra-osseous cysts.

Synovial Membrane

The outstanding features of the synovial membrane are hyperplasia, hypertrophy and increased vascularity. The thickened membrane falls into numerous folds and exhibits clusters of villous formations. (Fig. 17.)



Fig. 17. Note that the synovial membrane falls into folds and shows evidence of marked hyperplasia and hypertrophy and increased vascularity. Also, villus formations are readily discernible.

This is true of synovial membrane lining, the fibrous elements of the joint and the intra-articular fat pads. Deposits of blood pigment, varying in size, are readily discernible in the subsynovial layers. A marked increase in the fibrous tissue elements of the synovial membrane is readily demonstrable.

So far as one can determine from a clinical viewpoint, a hemophiliac is born with normal joints. However, the first hemorrhage into a joint may occur very early in childhood—the youngest patient with hemarthroses in this series was 12 months. However, most occur after 5 years of age. If a massive hemorrhage occurs in a joint that has not been implicated previously, or in a joint that has been the site of very few hemarthroses occurring at long intervals, the tissues return to normalcy without clinical manifestations of any unfavorable resid-

ual sequelae. After bleeding has been controlled, either spontaneously or by the administration of blood or plasma, the time interval required for absorption of the blood is governed by the massiveness of the hemorrhage and the joint involved. Large joints such as the knee and the shoulder may necessitate from 2 to 3 weeks before all clinical evidence of absorption of the blood is noted. In the joints under discussion one must assume that the synovial and the subsynovial tissues are able to handle the intra-articular blood with ease and not suffer any harmful effects in doing so. This is confirmed by clinical examination of these joints after all evidence of intra-articular blood has disappeared. These joints now are capable of full function and show no osseous or soft tissue alterations. Furthermore, it has been shown in experimental animals that injections of the knee joints by heparinized and nonheparinized blood at weekly intervals for from 4 to 6 weeks produced only a mild response in the synovial and the subsynovial tissues. It was noted also that if the animals were sacrificed 2 weeks after the last injection, the synovia returned to normalcy.

Repeated insults to the joint tissues by numerous and frequent hemorrhages, either of the massive or the subclinical variety, produce changes in all the joint elements that are irreversible and produce eventually fixed deformities and varying degrees of dysfunction consistent with chronic arthritis. The synovial and the subsynovial tissues are implicated early and show advanced alterations before the cartilaginous and the osseous elements are involved. The essential response of these tissues is a hyperplastic process producing a villous synovitis and thickening of the subsynovial tissues by marked fibrous tissue proliferation. These tissues are laden with blood pigment, leading many observers to conclude that this substance is the noxious agent responsible in a large measure for the hyperplastic response. Repeated pronounced intra-articular tension produced by numerous massive hemorrhages also must be regarded as a causative agent. That blood pigment is the irritant responsible for the alterations has not been confirmed by animal experimentation. It will be shown in a subsequent report that repeated massive injections of blood pigment extracted from the animals' own blood failed to produce alterations of the same intensity, as did a similar number of injections of heparinized or nonheparinized blood. This experiment forces one to conclude that some other agent in the blood may be the etiologic factor. Inasmuch as no severe alterations were observed when blood plasma was injected into the joints of another series of animals, one must look to some other factor in the blood as the irritant.

Regardless of the cause, the resulting hyperplastic process is a definite deterrent to resorption of blood from the joint cavity. Organization of the granulation tissue produces dense scar tissue eventually. A similar process is in progress in the fibrous capsule, the ligamentous apparatus and other periarticular tissues. Such changes provide an explanation of the clinical features noted in a joint that is the site of repeated hemarthroses If the knee joint is involved, one notes marked thickening of the parapatellar tissues. As a rule, soft crepitus is demonstrable on flexion and extension of the joint. Invariably such joints exhibit some restriction of motion. In more advanced cases contraction and shortening of the capsule and the surrounding muscles, particularly on the flexor side of the joint, result in fixed deformities. The hyperplastic synovial membrane at the peripheries of the articular cartilage forms a panus over the hyaline cartilage, depriving it of its nutrition In these areas rapid disintegration of the cartilage ensues and is invaded by vascular young connective tissue. From such areas, by metaplasia, cartilaginous or bony excrescencies are formed. These are observed in the gross specimens described previously and noted also in experimental animals.

The abnormalities in the articular cartilage have been recorded previously. As yet, the factor or the factors responsible for these changes are not known; some workers have suggested that some agent may exist in hemophilic blood which has a selective toxic effect on the articular cartilage. Such a factor has not been isolated as yet. The earliest changes in the cartilage comprise yellow staining and fibrillation of the superficial fibers; later, punched-out areas of destruction and erosion are noted. The entire surface appears pitted, while its margin is covered by a panus of synovial membrane that invades the degenerated cartilage. In such areas cartilaginous and bony excrescencies may form by metaplasia of the young connective tissue. The resulting irregular marginal excrescencies produce varying degrees of incongruity of the articulating surfaces of the bone ends of the affected joint. This feature is exhibited in the knee joints described in this presentation. On the other hand, gradual disintegration of the articular margins may occur, followed by resorption, so that the total area of the articular surface is reduced markedly; this abnormality is noted in the humeral head described previously. This specimen shows further that the entire cartilaginous surface may disappear, leaving behind a sclerotic, irregular, punched-out eburnated bony surface.

Cyst Formation

The cystic areas and clefts noted in the articular cartilage may arise from coalescence of Weichselbaum's lacunae, by replacement of deteriorated areas of articular cartilage by connective tissue or by the destruction of the overlying subchondral bone and cartilage by vascular connective tissue originating in the marrow spaces immediately adjacent to the subchondral bone. All these modes of origin are noted in the microscopic study recorded herein The proliferating connective tissue found in the cancellous bone appears to be a nonspecific response produced by death of cartilage cells and disintegration of the hyaline matrix. It was noted previously that this tissue also might cause resorption of the surrounding bony trabecluae, permitting many marrow spaces to communicate with one another, thereby producing cystic defects in the subchondral bone or in the cancellous bone. If the process progresses toward the joint cavity destroying the overlying cartilage in its path, the cysts will communicate eventually with the joint cavity.

The other theory for the formation of cysts is intra-osseous hemorrhage that breaks down the atrophic bony trabeculae found in the cancellous bone of the hemophiliac. Some cysts attain giant proportion. Coalescence and collapse of these defects produce marked deformities of the bone ends and incongruity of the articular surfaces. This feature, plus a reparative process in the quiescent stage, provides an explanation of the bony sclerosis observed roentgenologically in some of the joints. That healing may occur in cystic areas has been confirmed by a study of serial roentgenograms of joints over a period of several years. Occasionally the walls of the cysts become dense and sclerotic, precluding obliteration of the cyst. If the process continues to be active, sufficient fibrous tissue grows out of the denuded bone ends to permit development of fibrous ankylosis. Bony ankylosis does occur, but this is a rare sequela. The above alterations in the synovialis, the subsynovial tissue and the cartilaginous and the osseous elements, together with the change occurring in the capsule and the pericapsular tissue, are responsible for the establishment of fixed joint deformities of varying severity.

JOINT DEFORMITIES REQUIRING SPECIAL CONSIDERATION

The clinical and the roentgenologic features observed in some joints of the hemophiliac are of special interest because their development is peculiar to the joint implicated. This is particularly true of the knee, the elbow and the shoulder joints. Some of



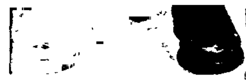


FIG. 18. Note the compound deformity of the right knee joint, comprising flexion of the joint, genu valgum and external rotation of the tibia.



FIG. 19. Observe the knobbiness of both knee joints. This is very characteristic in children after the age of 10 and in adults. This knobbiness is due to irregular overgrowth of the epiphyses and overgrowth of the patella.

the specific alterations are noted early in the development of the deformities, while others are observed in the late stages of development.

KNEE JOINT

It was recorded previously that this joint was the most frequent site of hemarthroses; also, it is usually the first joint to be implicated. The first intra-articular hemorrhage may occur in very early childhood. If repeated joint hemorrhages occur, the knee rapidly assumes a fixed deformity which it is difficult, and often impossible, to overcome. Such fixed deformities of varying severity have been observed in children under 5 years of age. The deformity is a compound one; it comprises flexion of the joint, genu valgum and external rotation of the tibia. (Fig. 18.) In long-established deformities, posterior subluxation of the tibia may exist. Once the tibia has subluxated posteriorly, it never returns to its normal position with or without treatment. It was recorded previously that the soft tissue alterations, especially in the young, played a very significant role in the development of the knee deformities.

Concurrent with contraction of the joint capsule, shortening and contracture of the hamstring muscles and the ilio-tibial band occur. These structures not only ensure a fixed flexion deformity but also produce external rotation of the tibia on the femur, the genu valgum, and in severe forms of this deformity, posterior subluxation of the tibia. It is interesting to note that in children exhibiting such deformities, the articular surface of the joint may show roentgenologically minimal or no alterations.

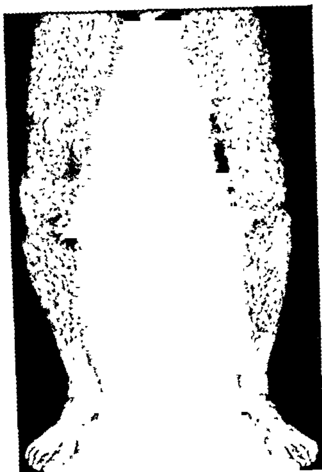


Fig. 20. Pronounced overgrowth of the medial one half of the proximal epiphysis of the tibia results frequently in a varus deformity of the tibia, as noted here. This is not an uncommon finding in adults.



Fig. 21. Note the overgrowth of the epiphyses and the epicondyles of the humerus and of the radial head of the right elbow joint in comparison with the left. Also observe the increase in size of the radial epiphysis of the right joint.

In children after the age of 10 and in adults the knee joints exhibit usually a characteristic knobbiness resulting from irregular overgrowth of the epiphyses; the patella may show marked overgrowth. (Fig. 19.) In some instances the epiphyses on the medial side of the joint show more overgrowth than those on the lateral side, so that the patella may be forced to assume a subluxated position on the lateral femoral condyle. Pronounced overgrowth of the medial one half of the proximal epiphysis of the tibia frequently produces a varus deformity of the tibia immediately below the medial tibial condyle; also, the medial one half of the distal femoral epiphysis may be enlarged This deformity is encountered most often in adults; usually it is bilateral. (Fig. 20.)

Elbow Joint

The features peculiar to the deformity of this joint are restriction of extension, varying degrees of valgity and knobbiness, and broadening of the epicondyles of the humerus. (Fig. 21.) The alterations are produced chiefly by irregular overgrowth of the epiphyses and in some instances by premature closure of the lateral one half of the distal epiphyseal plate of the humerus. Other interesting abnormalities encountered frequently in the elbow are excessive overgrowth and flaring of the ossification center comprising the head of the radius, premature closure of the epiphyseal plates of the secondary ossification centers such as those of the epicondyles, and pointing of the tip of the olecranon.

Shoulder Joint

Roentgenologically this joint frequently exhibits 2 interesting abnormalities. The



FIG. 22. Observe the large flowing beaklike osteophyte that forms frequently on the inferior articular margin of the head of the humerus in a hemophiliac.



FIG. 23. The superior articular surface of the talus is flattened, the bone is sclerotic, and the appearance of the talus is similar to that observed in osteochondritis.

neck of the scapulae may be elongated, and in some instances it is directed slightly downward. Large flowing beaklike osteophytes may form along the inferior articular margin of the head of the humerus. (Fig. 22.)

ANKLE JOINT

The most significant alteration in the ankle joint comprises flattening of the superior articular superior of the talus, which resembles the end-result of an osteochondritic process. (Fig. 23.) Similar changes are noted occasionally in the head of the femur; here the changes simulate Calvé-Perthes' disease.

CLASSIFICATION OF HEMOPHILIC ARTHRITIS

König classified the different stages of hemophilic arthropathy as (1) hemarthrosis, (2) panarthritis and (3) the regressive stage. Key does not recognize any significant differences between König's stage of panarthritis and the regressive stage; he described two types—(1) acute hemarthrosis and (2) chronic arthritis. Although Key's classification is more simple than that of Konig, it fails to recognize the progressive pathologic stages of the disorder, which in most instances parallels the clinical and the roentgenologic features. On the other hand, König's classification fails to correlate the clinical and the roentgenologic features with the clinical findings in terms of joint dysfunction, if present. Therefore, in this study an attempt was made to classify hemophilic arthropathy into 4 grades which correlate the clinical manifestations with the pathologic alterations in the components of the joint and with the observations noted roentgenologically.

Acute hemarthrosis is not included in this classification because it may occur in a normal joint or in a joint showing alterations of varying severity. The response of the joint tissues to massive intra-articular bleeding depends upon the changes existing in the joint prior to the bleeding. If bleeding occurs in a normal joint, as a rule the tissues return to normalcy within a few days. However, repeated hemorrhages of this nature produce permanent alterations which tend to dispose the joint to future episodes of bleeding which, in turn, increase the severity of the existing tissue changes. Acute

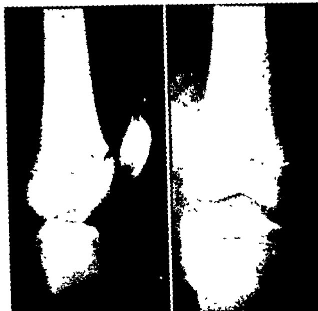


FIG. 24. This knee joint is an example of the changes designated Grade 1. Note that there are no osseous changes. However, there is some thickening of the synovial membrane and the subsynovial tissues, as evidenced by the increased density of the soft tissues surrounding the joint.



FIG. 25. This is an example of the alterations designated Grade 2. Note that the bony trabeculae of the epiphyses appear to be stouter than normal and stand out in bold relief. However, there is no incongruity of the joint surfaces, and the joint spaces are well preserved. No defects are noted in the articular cartilage, and no cysts are noted in the subchondral bone.

hemarthrosis in joints afflicted with chronic arthritis behaves differently from that occurring in normal or nearly normal joints. In the former, the blood is absorbed more slowly because the alterations in the synovial, the subsynovial and the capsular tissues preclude rapid dispersion and absorption of the blood. The blood and the resulting increased intra-articular tension in such joints enhance formation of fibrous tissue in the soft tissue elements and the alterations described previously in the cartilaginous and the osseous components of the joint. It becomes apparent that acute hemarthrosis must be considered as an important factor in initiating arthritis and increasing its severity, but it should not be considered per se as a type of arthritis. In this study the disorder has been divided into Grades 1, 2, 3 and 4.

GRADE 1

This is the earliest form of arthritis encountered. Essentially the changes are limited to the soft tissue elements. Clinically the joint exhibits no restriction in its arcs of motion, and no deformities are discernible. Slight atrophy of the muscles about the joint may be present; this is especially true of the knee joint. Some thickening of the synovial and the subsynovial tissues is discernible, but as yet no broadening of the epiphysis has occurred. Roentgenograms exhibit no bony abnormalities, except for some slight generalized decreased density of the bone ends. However, they may reveal some increased density of the capsular tissues. At this stage the joint exhibits no dysfunction. (Fig. 24.)

GRADE 2

In this stage all the observations noted in Grade 1 are more pronounced. In addition, deformities of joints in one plane are discernible. No angular deformities are present. The deformities are due entirely to contractures implicating the capsular and the pericapsular tissues, resulting in loss of a few



FIG. 26. *This is an example of the alterations designated* Grade 3. Observe the marked atrophy of the muscles, pronounced pericapsular thickening of the joints with some knobbiness and the compound deformity at the knee joint comprising flexion, genu valgum and external rotation of the lower leg.

degrees of the normal arcs of motion. However, this *restriction of motion does not* produce serious impairment of the over-all performance of the joint, and the patients are not aware of any appreciable dysfunction. *Usually the bone ends exhibit slight* broadening, and the first indications in a mild form of knobbiness of the joint are manifest. Roentgenologic study reveals no incongruity of the joint surfaces, and the joint space is well preserved. No defects in the articular cartilage or the cystic formation in the subchondral bone or cancellous bone are noted. However, the bony trabeculae of the epiphysis may appear stouter than normal and stand out in bold relief. (Fig. 25.)

GRADE 3

Joints in this stage of development of the arthritic process show fixed deformities which in some instances are in more than one plane. Examples of this grade are readily demonstrable in the knee, the elbow and the subtalar joints. The knee may exhibit flexion, valgity and external rotation of the tibia on the femur. Occasionally a varus deformity may be present. The elbow may exhibit varying degrees of valgity, and the foot may be everted at the subtalar joint, resulting from contracture of the peroneal muscle mass The combined soft tissue and cartilaginous and osseous changes are of sufficient severity to produce an appreciable degree of dysfunction. In addition, clinical examination reveals marked atrophy of the



FIG. 27. This is the roentgenogram of the patient depicted in Figure 26 Note thinning of the joint space, beginning spur formation, particularly of the tibial spines, punched-out cysts in the subchondral areas, particularly of the tibia, and incongruity of the articular surfaces



FIG. 28. These alterations are consistent with Grade 4. Note the pronounced thickening of the periarticular tissues and the knobbiness of the knee joint.



FIG. 29. These roentgenograms are of the person depicted in Figure 28. Note the marked thinning of the joint spaces, incongruity of the articular surfaces, spur formation and subchondral cyst formation.

muscles motorizing the joint, pronounced pericapsular and capsular thickening, and knobbiness of the bone ends. (Fig. 26.) Roentgenologically one notes irregular overgrowth of the epiphysis, incongruity of the articular surfaces, clefts and punched-out areas in the cartilage, cysts in the subchondral and the cancellous bone, slight thinning of the joint space and beginning spur formations. (Fig. 27.) Bone sclerosis is not a significant feature of this stage. These joints often become painful and stiff with activity incident to normal function and often are the site of repeated massive and subclinical hemorrhages.

GRADE 4

This stage embraces the most severe alterations encountered in this study. Here the changes noted in Grade 3 are more pronounced, and the functional capacity of the joint is reduced greatly. Frequently, soft crepitus is demonstrable in the joint while performing within its restricted arcs of motion. The deformities are fixed and pronounced. Marked knobbiness is a constant feature, and in some instances fibrous ankylosis exists. Fibrous ankylosis is a frequent finding in the patellofemoral joint. In many of these joints the patella is fixed firmly to the anterior surface of the femur, and occasionally it assumes a subluxated position. Roentgenologic study reveals bony sclerosis to be the outstanding feature. In addition, part or all of the joint space is thinned or obliterated completely. Marginal spurring and spurring of bony processes such as the olecranon and the tibial spines are concomitant findings in most joints in this category. Cysts in varying steps of development and in some cases in stages of healing are discernible. It becomes apparent that pronounced incongruity of the articular surfaces is a constant observation. It is interesting to note that these alterations generally are noted in adults in whom the frequency of joint hemorrhages is reduced greatly. It appears that once the arthritic process is initiated and progresses to a level of severity sufficient to alter the normal joint mechanics, progression of the alterations continue in spite of the infrequency or the absence of acute hemarthroses. (Figs. 28 & 29.)

MANAGEMENT OF THE HEMOPHILIAC

PROPHYLAXIS

Generally it is accepted that in most instances trauma—minor or severe—initiates

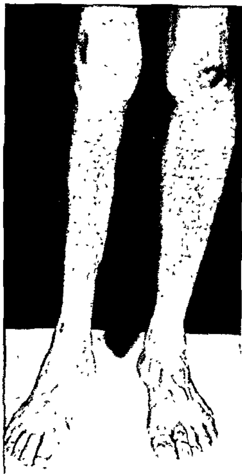


FIG. 26. This is an example of the alterations designated Grade 3. Observe the marked atrophy of the muscles, pronounced pericapsular thickening of the joints with some knobbiness and the compound deformity at the knee joint comprising flexion, genu valgum and external rotation of the lower leg.

degrees of the normal arcs of motion. However, this restriction of motion does not produce serious impairment of the over-all performance of the joint, and the patients are not aware of any appreciable dysfunction. Usually the bone ends exhibit slight broadening, and the first indications in a mild form of knobbiness of the joint are manifest. Roentgenologic study reveals no incongruity of the joint surfaces, and the joint space is well preserved. No defects in the articular cartilage or the cystic formation in the subchondral bone or cancellous bone are noted. However, the bony trabeculae of the epiphysis may appear stouter than normal and stand out in bold relief. (Fig. 25.)

GRADE 3

Joints in this stage of development of the arthritic process show fixed deformities which in some instances are in more than one plane. Examples of this grade are readily demonstrable in the knee, the elbow and the subtalar joints. The knee may exhibit flexion, valgity and external rotation of the tibia on the femur. Occasionally a varus deformity may be present. The elbow may exhibit varying degrees of valgity, and the foot may be everted at the subtalar joint, resulting from contracture of the peroneal muscle mass The combined soft tissue and cartilaginous and osseous changes are of sufficient severity to produce an appreciable degree of dysfunction. In addition, clinical examination reveals marked atrophy of the



FIG. 27. This is the roentgenogram of the patient depicted in Figure 26. Note thinning of the joint space, beginning spur formation, particularly of the tibial spines, punched-out cysts in the subchondral areas, particularly of the tibia, and incongruity of the articular surfaces.



FIG. 28. These alterations are consistent with Grade 4. Note the pronounced thickening of the periarticular tissues and the knobbiness of the knee joint.



FIG. 29. These roentgenograms are of the person depicted in Figure 28. Note the marked thinning of the joint spaces, incongruity of the articular surfaces, spur formation and subchondral cyst formation.

muscles motorizing the joint, pronounced pericapsular and capsular thickening, and knobbiness of the bone ends. (Fig. 26.) Roentgenologically one notes irregular overgrowth of the epiphysis, incongruity of the articular surfaces, clefts and punched-out areas in the cartilage, cysts in the subchondral and the cancellous bone, slight thinning of the joint space and beginning spur formations (Fig. 27.) Bone sclerosis is not a significant feature of this stage. These joints often become painful and stiff with activity incident to normal function and often are the site of repeated massive and subclinical hemorrhages.

GRADE 4

This stage embraces the most severe alterations encountered in this study. Here the changes noted in Grade 3 are more pronounced, and the functional capacity of the joint is reduced greatly. Frequently, soft crepitus is demonstrable in the joint while performing within its restricted arcs of motion. The deformities are fixed and pronounced. Marked knobbiness is a constant feature, and in some instances fibrous ankylosis exists. Fibrous ankylosis is a frequent finding in the patellofemoral joint. In many of these joints the patella is fixed firmly to the anterior surface of the femur, and occasionally it assumes a subluxated position. Roentgenologic study reveals bony sclerosis to be the outstanding feature. In addition, part or all of the joint space is thinned or obliterated completely. Marginal spurring and spurring of bony processes such as the olecranon and the tibial spines are concomitant findings in most joints in this category. Cysts in varying steps of development and in some cases in stages of healing are discernible. It becomes apparent that pronounced incongruity of the articular surfaces is a constant observation It is interesting to note that these alterations generally are noted in adults in whom the frequency of joint hemorrhages is reduced greatly. It appears that once the arthritic process is initiated and progresses to a level of severity sufficient to alter the normal joint mechanics, progression of the alterations continue in spite of the infrequency or the absence of acute hemarthroses. (Figs. 28 & 29.)

MANAGEMENT OF THE HEMOPHILIAC

PROPHYLAXIS

Generally it is accepted that in most instances trauma—minor or severe—*initiates*

hemarthroses. It becomes apparent that parents should handle hemophilic infants with gentleness and care. The activities of children should be curtailed in order to protect them from abnormal stress and trauma. However, it is difficult to achieve this, particularly when children are sent to school and are not under the close supervision of their parents. Older children must be informed concerning their affliction and how vital it is for them to avoid bodily injury. Adults have fewer hemorrhages because they learn to live with their affliction and protect their bodies from excessive activity and trauma.

Many acute hemarthroses begin slowly and reach large proportions only if they are ignored. Parents should be taught to recognize early bleeding. Usually this is associated with pain, stiffness and restricted activity due to muscle spasm. At this stage the joint should be put at complete rest and, where possible, a compression bandage applied; then the part is surrounded by ice packs. *If these precautions are taken, many* large hemarthroses will be aborted.

Massive Hemarthrosis

This may occur suddenly or insidiously; the management should be directed concurrently to the local lesion and the blood defect. In order to treat these patients adequately, centers should be set up where the patients are surveyed periodically and they can report and be admitted in case of emergency. At these centers, blood banks with sufficient blood of all types to meet the emergency should be readily available; also, frozen plasma should be kept in stock to be used in case the specific type of blood is not on hand. Such a center has been instituted at Jefferson and has serviced many of the hemophiliacs with all forms of acute hemorrhages

Patients with acute massive hemarthroses or with mild hemarthroses that do not respond to the measures described previously are admitted to the hospital and put at complete bed rest. The blood defect is studied immediately and corrected by the administration of whole blood or plasma. Transfusions are given repeatedly, even after the bleeding time has approached normal limits. This is an important facet in the management of acute hemorrhages, because patients may continue to bleed in spite of a normal bleeding time and can be controlled only by repeated administration of normal blood.

The joint is put at complete rest and surrounded by ice packs. Where possible, an elastic compression bandage is applied. This is especially applicable to knee, ankle, elbow and wrist joints. The best way to attain uniform compression is first to lay on the anterior, the posterior and the lateral surfaces of the joint wide strips of foam rubber ½ in. thick; these extend 2 or 3 in. proximal and distal to the joint; then the extremity with the strips in place is wrapped with several layers of elastic bandage. This dressing serves as a means of compression and also immobilizes the joint. Ice packs are applied over the compression dressing.

At this point the advisability of aspiration of the joint or the instillation of some dispersing agent into the joint must be considered. Increased intra-articular tension and the blood per se are noxious agents capable of producing deleterious effects in the joint tissues. Therefore, it becomes obvious that reduction of the tension and removal of the blood are desirable in acute hemarthroses. Whenever feasible, aspiration of blood is performed routinely in this clinic; in fact, if necessary, repeated aspirations are done. This is followed by the application of a compression bandage such as described previously As yet, no unfavorable sequelae have followed joint aspirations It is felt that aspirations never are contraindicated, provided proper measures are taken to control the blood defect

The authors are of the opinion that spreading factors such as hyaluronidase should not be employed in massive intra-articular hemorrhage. This is based on the

clinical observation that joints treated by such spreading agents return to normalcy much slower than do joints treated by aspiration and compression. In the former joints, pronounced diffuse thickening of the synovial and the capsular tissues is demonstrable for many weeks after the acute hemorrhage. One knee joint treated by the injection of hyaluronidase disclosed marked painful thickening of tissues 7 weeks after the injection. Hyaluronidase causes depolymerization of the principal element of the ground substance, namely, the mucopolysaccharide hyaluronic acid, and increases the permeability of the synovial membrane. These conditions favor rapid absorption of intra-articular blood, particularly when under great tension. However, when this occurs, the synovial membrane is forced to carry a tremendous work load within a brief period of time and is subjected suddenly to a concentration of noxious factors in the blood. Clinical evidence of this conclusion is borne out by the marked thickening of the synovial tissue following the injection of the enzyme into a site of hemarthrosis and by the prolonged period that it takes the tissues to return to normalcy or near normalcy. It becomes apparent that repeated insults of this nature have serious deleterious effects on the joint tissues and will enhance development of arthritic alterations. When hyaluronidase first was employed in this clinic as an adjunct in the treatment of hemarthrosis, the authors were impressed by the immediate favorable response of the joints and even recommended its use, but as the clinical material accumulated and a critical analysis was made of the cases treated by the 2 methods in the follow-up clinic, they were forced to change their first impression. However, they do believe that hyaluronidase has a place in the treatment of massive hemorrhages into soft tissues such as muscles, subcutaneous tissue and skin. Early injection of the enzyme promotes rapid absorption of the blood and precludes thrombosis of vessels and necrosis of tissue.

After aspiration of the joint, just as soon as it becomes apparent that bleeding is controlled, active motion within the painless arcs is instituted. However, no weight-bearing is permitted at this time. Active motion favors absorption of any residual blood, enhances nutrition of the articular cartilage and prevents contracture of capsular and pericapsular tissues. Active motion may be performed within the compression dressing. If the knee joint is implicated, care must be taken to prevent flexion deformity of the joint. This it achieved by traction to the lower leg. However, traction should not be applied if the knee is distended markedly, because the extended position causes the intra-articular tension to rise, thereby producing more pain and exerting more tension on the synovial tissues. Traction is applied after the blood is aspirated and bleeding is controlled.

The quadriceps apparatus should be given special consideration. Quadriceps atrophy and loss of power are constant concomitant findings following hemarthrosis of the knee joint. In some instances complete loss of voluntary contraction of the muscle ensues; in 1 case 12 weeks passed before even a flicker of contraction was demonstrable in the muscle. This occurred in spite of intensive efforts on the part of the patient to restore muscle power. It becomes apparent that early quadriceps setting and resistance exercises must be instituted just as soon as pain is alleviated and bleeding is controlled. The knee joint should be protected for several weeks by crutch-walking until the muscle is sufficiently strong to stabilize the knee joint adequately. It is wrong to allow patients to wear a long leg brace in order to prevent hemarthroses; this promotes greater atrophy of the quadriceps and favors stiffness of the knee joint. Long leg braces to be worn permanently are prescribed only for patients in whom the deformity is beyond correction



FIG. 30. This long leg brace with a circular spring is worn part of the day and during sleeping hours in an effort to prevent and correct flexion deformities of the knee joints in hemophiliacs.

and some sort of protection to the limb is needed.

CORRECTION OF JOINT DEFORMITIES

Every effort should be made to prevent the development of joint deformities, because once established they are very difficult to correct. The most effective preventive measure is early active motion within the tolerance of pain immediately after a hemarthrosis. This promotes absorption of intra-articular blood, enhances normal exchange of interstitial tissue fluids, minimizes fibrous tissue formation and contractures of ligaments, capsule, tendons and muscles. Deformities of the knee joint produce the greatest dysfunction, hence, deformities of this joint will be considered in detail.

Following acute hemarthroses, knee joints, especially in children, show a great tendency to develop fixed flexion deformities. If, after the measures described previously, complete correction is not maintained, a long leg brace with a circular spring at the knee joint is worn for part of the day and during the sleeping hours. (Fig. 30.) This spring is designed to exert continuous extension at the knee joint, yet it allows some voluntary flexion. While wearing the brace, resistance exercises are performed on a regulated regimen. The brace is discarded as soon as complete extension can be performed voluntarily and the tendency for development of a flexion deformity is overcome.

Mild flexion deformities that fail to respond to this method can be corrected by continuous skin traction or wedging plaster casts. Moderate and severe flexion deformities usually are associated with valgity at the knee joint and external rotation of the tibia on the femur. Regardless of the methods employed, the authors never have been able to correct the last 2 components of the deformity. Furthermore, flexion deformity in these joints may be corrected, but invariably the tibia assumes a position of posterior subluxation. Unfortunately, such deformities not only cause marked dysfunction but also predispose the joint to recurrent hemorrhage. Some of these patients are doomed to a long leg brace for life because there is nothing else to offer from a conservative point of view

Such hopeless cases have been the stimulus for investigating a surgical approach to this problem. The hazards associated with surgery on the hemophiliac are well known; nevertheless, the authors feel that in properly selected cases and with adequate preoperative and postoperative management, surgical correction of some of these deformities

is possible. In the selection of cases, patients with severe blood defects are excluded; only patients with mild or moderate defects of Grades 1 and 2 are considered suitable subjects.* The surgical technic should be such that no tissue planes are developed; no tissue spaces are left open, and closure is achieved by through-and-through interrupted sutures. Incisions are made with a cautery, and deep wounds, if not collapsible by external pressure, should be drained to prevent accumulation of blood. By observing these rules in this clinic the following surgical procedures have been performed without any unfavorable sequelae: 1 arthrodesis of the knee joint; 1 arthrodesis of the hip joint; 1 massive excision of necrotic tissue and replacement with skin grafts and 2 shelf operations and 2 derotation osteotomies for congenital dislocation of the hip joints. We state emphatically that surgery in the hemophiliac is dangerous; never should it be attempted except in those cases in which all other means have been exhausted, and then only if the patient's blood defect is of Grade 1 or 2 and if the personnel and the physical equipment are available to administer adequate preoperative and postoperative management.

* Dr. L. Tocantins uses 4 grades in classifying hemophiliacs, the grade depending on the severity of the blood defect. This classification and the criteria of each grade will be released for publication in the next few months.

BIBLIOGRAPHY

Caffey, J., and Schlesinger, E. R.: Certain effects of hemophilia on growing skeleton: some roentgenographic observations on overgrowth and dysgenesis of epiphyses associated with hemarthrosis, J. Pediat. 16:549-565, 1940.

Doub, H. P., and Davidson, E. C.: Roentgenray examination of the joints of hemophiliacs, Radiology 6:217, 1926.

Freund, E.: Die Gelenkerkrankungen der Bluter, Arch. path. Anat. 256:158-189, 1925.

Johnson, J. B., Davis, T. W., and Bullock, W. H.: Bone and joint changes in hemophilia, Radiology 63:64-71, 1954.

Key, J. A.: Hemophilic arthritis, Ann. Surg. 95:198, 1932.

König, Franz: Die Gelenkerkrankungen bei Blutern mit besonderer Berücksichtigung der Diagnose, Klin. Vorträge, N. F. No 36 (Chirurgie No. 11) pp. 233-243, 1892.

Reinecke and Wohlwill: Hemophilic arthritis, Arch. klin. Chir. 154:425, 1929.

Thomas, H. B.: Some orthopaedic findings in 98 cases of hemophilia, J. Bone & Joint Surg. 18:140-147, 1936.

Arthropathia Hemophilic

Summario in Interlingua

Le presente studio es basate super un analyse clinic del membros de 117 familias hemophilic studiate al Hospital del Collegio Medical Jefferson. Le studio resultava in observationes clinic relative al factores que es responsabile pro le deformitates del articulation. Iste factores es dolor e spasmos muscular resultante ab massive hemorrhagias e—plus importantemente—repetite minor hemorrhagias a in le articulationes afficite. Deformitates articulatori pote etiam esser le resultato de massive hemorrhagias a in le musculo e le plano fascial. Exemplos de tal deformitates es demonstrate per un caso resimilante contractura de Volkman e un altere caso de contractura adductional del pollice. In certe casos massive hemorrhagias pote causar le destruction total del histos superjacente del articulation con le resultante necessitate de excisiones del histos necrotic sequite per graffos cutanee. Le altere factores responsabile es embalancia muscular, alterationes del cartilagine articular, alterationes del osso subchondral, e alterationes hyperplastic del elementos de histos molle del articulationes. In le presente studio le aspectos roentgenographic del disordine es registrate. Iste aspectos include alterationes

observate in le elementos de histo molle e in le elementos cartilaginose e ossee del articulation. Attention special es prestate al characteristicas del formation de cystes subchondral le quales pote esser de varie dimensiones—usque a dimensiones gigante. Es etiam sublineate le effecto de arthropathia hemophilic super le crescentia del epiphyses. In certe casos il ha excessos de crescentia del epiphyses del articulation afficite. Iste aspecto es etiam responsabile, a un certe grado, pro le observate deformitates articulational. Es etiam listate le constatationes microscopic. Istos non differe ab le constatationes facite in altere typos de arthritis, specialmente in osteoarthritis. Es monstrate specimens obtenite post morte. Illos veni ab duo articulationes del humero e ab duo articulationes del genu. Es presentate le characteristicas micro- e macroscopic. Finalmente le tractamento de arthropathia hemophilic es delineate con emphase special super le prevention de deformitates quandocunque isto es possibile. Le arthropathia ha essite subdividite in plure gruppos clinic e, intra iste gruppos, in typos secundo le specific defecto sanguinee involvite. Esseva constatate que multes de iste patientes qui representa typos 1 e 2 pote esser subjicite a un tractamento chirurgic sin effectos adverse. Isto da al chirurgo le privilegio de corriger certes inter iste deformitates per methodos chirurgic. Tamen, le intervention chirurgic debe remaner le ultime recurso in le tractamento de arthropathia hemophilic.

16

Scapulocostal Syndrome*

RICHARD B. MCGOVNEY, M.D.†

The scapulocostal syndrome is a name given by A. A. Michele[2] in 1950 to a symptom complex involving unilaterally the shoulder and the region of the superior scapula with radiation of pain patterns into the neck and the head, the deltoid muscle, the arm and the hand, and occasionally the upper anterior chest wall. This is a common, painful condition encountered more frequently between the ages of 30 and 60, equally divided between the sexes, usually chronic at the time of diagnosis and amenable to treatment in a high percentage of cases. It is not to be confused with numerous other painful conditions involving the neck, the shoulder girdle and the upper extremity, such as cervical osteoarthritis, bursitis, herniated cervical intervertebral disk, brachial plexus neuritis, "rotator cuff" injuries, myositis and angina pectoris, although it may be a part of all of these. It has been stated variously that it is the cause of from 30 to 50 per cent of all unilateral pains in the neck, the shoulder and the arm areas in middle-aged persons.

There appear to be 2 main types—the atraumatic and the traumatic. The atraumatic is characterized by a more or less insidious onset from 30 to 60 years of age, with postural and/or occupational tension stresses without history of recent pertinent trauma to the involved regions. The traumatic type is characterized by a more acute or earlier onset of local or reflex symptoms following a history of injury to the neck, the shoulder or any part of the upper extremity. Russek[4] has classified these as primary, secondary and static. I feel that these 2 general types may coalesce or be superimposed, in that the potential for the atraumatic type may be present, and trauma may intervene as a precipitant.

History and examination of a patient with the atraumatic type usually will reveal a person between 30 and 65 who complains of unilateral pain in the head, the neck, the posterior shoulder, the arm and the hand, or various combinations of these, of several months' duration without apparent cause, with malpostural tensions, no obvious restrictions of motion or atrophy or neurovascular disturbances and an acute trigger point about the superior vertebral angle of the scapula. The traumatic type usually will be encountered in a somewhat younger person, on the average; the same symptom distribution will prevail, but there will have been within a fairly recent period some injury to the neck, the shoulder or the upper extremity causing voluntary or involuntary restriction of motion in these regions, with resulting loss of motion in any of the joints and the concomitant associated objective clinical findings: there also will be an acute trigger point about the superior vertebral angle of the scapula or the base of the spinous process of the scapula.

What is the etiology of this syndrome? To

---

* Paper presented at the meeting of the Western Orthopedic Association held in Hawaii, November, 1955.

† Santa Barbara, Calif.

my mind it has not been entirely established. Along with Michele, I postulate that it is a complex, the result of a fascial contraction —nerve irritation—segmental nerve reflex. As early as 1922 Henry J. Prentiss,[3] Professor of Anatomy at the University of Iowa, taught that

> the prevertebral fascia extends from the anterior tubercles of one side to the anterior tubercles of the other side, enclosing the prevertebral muscles with lateral extension of this fascia from the anterior tubercles of the cervical vertebrae around to the ligamentum nuchae and the spinous processes of the vertebrae—beneath it are all the lateral and postvertebral groups of muscles—all the spinal nerve trunks in the neck pass through this fascia.

He taught further that

> surrounding both the anterior and posterior compartments of the neck is the vaginal fascia (fascia profunda). This fascia is part of the great deep fascia which covers the whole body and separates the deeper structures from the superficial fascia and the skin and is continuous by septa with all the deeper structures and is not loosely applied like the superficial fascia.

Prentiss also said that

> this muscle fascia of the neck is continued down beneath the clavicle into the axilla as the costocoracoid membrane and is anchored to the clavicle by a reflection around the subclavius muscle

To understand this fascial-nerve-reflex concept we must understand these anatomic facts—that all the spinal nerve trunks in the neck pass through this fascia, and that this fascia is a part of the fascia profunda that covers the whole body and is continuous by septa with all the deeper structures. This latter includes, of course, the intermuscular and the intramuscular septa, points of bony attachment and neurovascular bundles.

Why are the trigger points in approximately 86 per cent of the cases located at or about the superior vertebral angle of the scapula? To my mind, this point also is not explained fully. I postulate that it is because this area of the scapula sustains the greatest suspensory or drag strain on the cervical fascia and muscles through which pass the spinal nerves involved in these pain patterns, and that cord segmental reflex symptoms are referred here.

Of the 3 bones that comprise the shoulder girdle, the scapula is the only one that articulates with the other 2—the common

---

TOTAL NUMBER OF PATIENTS: 150

Females 78 (52%)		Males 72 (48%)
	Side Involved	
R 95 (63.3%)	FR 50 (33.3%)	MR 45 (30%)
L 55 (36.7%)	FL 28 (18.66%)	ML 27 (18%)
Females R 64.1%		Males R 62.5%
L 35.9%		L 37.5%

There were 78 females, 72 males—essentially equal. With regard to the side involved, the figures show a very clear percentile occurrence between the sexes *as to area. Of the females, 64.1 per cent* were right, 35.9 per cent were left; of the males, 62.5 per cent were right, 37.5 per cent were left.

AVERAGE AGE: 45.09

Females 46.19	Males 44.00
30-40	40-50
F 18 (12.0%)	F 21 (14%)
M 17 (11.3%)	M 15 (10%)
50-60	60-70
F 12 (8%)	F 13 (8.66%)
M 15 (10%)	M 10 (6.66%)
10-20	20-30
F 2 (1.33%)	F 7 (4.66%)
M 3 (2.0%)	M 5 (3.33%)
70-80	80+
F 5 (3.33%)	F 2 (1.33%)
M 1 (0.66%)	M 0
Oldest F 87	Youngest F 14

The average age was 45.09 years, or the middle of the 5th decade. The average for females was 46.19 years, for males, 44 years. Numbers and percentages are given for the various decades from the 2nd to the 8th. They show that 80.66 per cent were in the age groups between 30 and 70. Of these, 64, or 52 89 per cent, were females, and 57, or 47.11 per cent, were males. The oldest patient was a female aged 87; the youngest, a female aged 14 years.

denominator. Of the multitude of muscles acting on the shoulder girdle, the deltoid is the only muscle that has bony attachments to each of the 3 bones, and it is in the deltoid alone that 44 per cent of referred pain in this syndrome occurs and in 32 per cent in addition in combination with the neck and the arm, making 76 per cent in which the deltoid area is involved. By far the greatest area of deltoid bony attachment is to the scapula. The one common tissue that applies to and acts on all other deep structures of neck, scapula, clavicle, humerus, spinal nerves, vascular bundles, muscles and intermuscular septa is the fascia profunda.

Fascia is a dense fibrous connective tissue that has the property of slight stretching and contracting particularly when a long-continued force is applied, and in this process stretches or constricts the structures that pass through or are attached to it. One important tissue passing through fascia is the spinal nerve that carries the afferent and the efferent impulses, which, when from an irritative source, are interpreted as pain.

The scapula is a suspended bone except for its articulation with the clavicle, which in itself is a bone suspended by soft tissues having the same or closely similar cord segmental innervations as do the muscles and fascia suspending the scapula. The muscles and the fascias that suspend the scapula all originate in the cervical or upper dorsal vertebrae, or are extension of the prevertebral and the postvertebral fascias that are penetrated by the spinal nerves Any drag or sag exerted by the scapula in abnormal postural positions puts stress on these muscles and fascias, which in turn stimulate the penetrating nerves and initiate painful stimuli. Inman, Saunders and Abbott[1] state that motion occurs simultaneously in all joints of the region (shoulder girdle), each contributing its share. At the glenohumeral and the scapulothoracic articulations the ratio from almost the beginning to the termination of the arc is respectively 2 to 1. Steindler[5] states that abduction in the glenohumeral joint is associated from the beginning with scapular rotation.

The region of the scapula that has the greatest degree of motion, either translatory (that is, forward or back, up or down) or rotary, and also is supplied by the same cord segmental areas as the neck, the shoulder girdle and the upper extremity, is the area about the superior vertebral angle.

Tarsy[6] states that the spread of pain is

Duration

5 Yr.	9	39, or 26%
2 Yr.	10	
1 Yr.	20	
6 Mo.	13	110, or 74%
3 Mo.	19	
1 Mo.	29	
1 Wk.	34	
1 Day	15	
Unknown	1	

More than 1 Yr.	26%
1 Yr. or less	87.3%
Less than 6 Mo.	74%
Less than 1 Mo.	32%

The duration of symptoms varied from over 5 years to 1 day. Those lasting more than 1 year were 26 per cent; 1 year or less, 87.3 per cent; less than 6 months, 74 per cent; and less than 1 month, 32 per cent. The average duration approximated 8 months.

Symptom Area

	Cases
Shoulder	66 (44%)
Shoulder and Neck	26 (17.3%)
Scapula	23 (15.3%)
Shoulder and Arm	22 (14.7%)
Arm and Hand	19 (12.7%)
Neck and Head	15 (7.3%)
Neck and Arm	2 (1.3%)
Bicipital Groove	2 (1.3%)
Chest	2 (1.3%)
Elbow	1 (0.67%)

The symptom area is shown. The shoulder was involved in 114, or 76 per cent, and, with the scapula, in 137, or 91.3 per cent, of the cases.

my mind it has not been entirely established. Along with Michele, I postulate that it is a complex, the result of a fascial contraction —nerve irritation—segmental nerve reflex. As early as 1922 Henry J. Prentiss,[3] Professor of Anatomy at the University of Iowa, taught that

> the prevertebral fascia extends from the anterior tubercles of one side to the anterior tubercles of the other side, enclosing the prevertebral muscles with lateral extension of this fascia from the anterior tubercles of the cervical vertebrae around to the ligamentum nuchae and the spinous processes of the vertebrae—beneath it are all the lateral and postvertebral groups of muscles—all the spinal nerve trunks in the neck pass through this fascia.

He taught further that

> surrounding both the anterior and posterior compartments of the neck is the vaginal fascia (fascia profunda). This fascia is part of the great deep fascia which covers the whole body and separates the deeper structures from the superficial fascia and the skin and is continuous by septa with all the deeper structures and is not loosely applied like the superficial fascia.

Prentiss also said that

> this muscle fascia of the neck is continued down beneath the clavicle into the axilla as the costocoracoid membrane and is anchored to the clavicle by a reflection around the subclavius muscle

To understand this fascial-nerve-reflex concept we must understand these anatomic facts—that all the spinal nerve trunks in the neck pass through this fascia, and that this fascia is a part of the fascia profunda that covers the whole body and is continuous by septa with all the deeper structures. This latter includes, of course, the intermuscular and the intramuscular septa, points of bony attachment and neurovascular bundles.

Why are the trigger points in approximately 86 per cent of the cases located at or about the superior vertebral angle of the scapula? To my mind, this point also is not explained fully. I postulate that it is because this area of the scapula sustains the greatest suspensory or drag strain on the cervical fascia and muscles through which pass the spinal nerves involved in these pain patterns, and that cord segmental reflex symptoms are referred here.

Of the 3 bones that comprise the shoulder girdle, the scapula is the only one that articulates with the other 2—the common

TOTAL NUMBER OF PATIENTS: 150

Females 78 (52%)		Males 72 (48%)
	Side Involved	
R 95 (63.3%)	FR 50 (33.3%)	MR 45 (30%)
L 55 (36.7%)	FL 28 (18.66%)	ML 27 (18%)
Females R 64.1%		Males R 62.5%
L 35.9%		L 37.5%

There were 78 females, 72 males—essentially equal. With regard to the side involved, the figures show a very clear percentile occurrence between the sexes as to area. Of the females, 64.1 per cent were right, 35.9 per cent were left; of the males, 62.5 per cent were right, 37.5 per cent were left.

AVERAGE AGE: 45.09

Females 46.19	Males 44 00
30-40	40-50
F 18 (12.0%)	F 21 (14%)
M 17 (11.3%)	M 15 (10%)
50-60	60-70
F 12 (8%)	F 13 (8.66%)
M 15 (10%)	M 10 (6 66%)
10-20	20-30
F 2 (1 33%)	F 7 (4.66%)
M 3 (2.0%)	M 5 (3.33%)
70-80	80+
F 5 (3.33%)	F 2 (1.33%)
M 1 (0.66%)	M 0
Oldest F 87	Youngest F 14

The average age was 45.09 years, or the middle of the 5th decade. The average for females was 46.19 years, for males, 44 years Numbers and percentages are given for the various decades from the 2nd to the 8th. They show that 80 66 per cent were in the age groups between 30 and 70. Of these, 64, or 52.89 per cent, were females, and 57, or 47.11 per cent, were males. The oldest patient was a female aged 87; the youngest, a female aged 14 years.

denominator. Of the multitude of muscles acting on the shoulder girdle, the deltoid is the only muscle that has bony attachments to each of the 3 bones, and it is in the deltoid alone that 44 per cent of referred pain in this syndrome occurs and in 32 per cent in addition in combination with the neck and the arm, making 76 per cent in which the deltoid area is involved. By far the greatest area of deltoid bony attachment is to the scapula. The one common tissue that applies to and acts on all other deep structures of neck, scapula, clavicle, humerus, spinal nerves, vascular bundles, muscles and intermuscular septa is the fascia profunda.

Duration

5 Yr.	9	39, or 26%
2 Yr.	10	
1 Yr.	20	
6 Mo.	13	110, or 74%
3 Mo.	19	
1 Mo.	29	
1 Wk.	34	
1 Day	15	
Unknown	1	

More than 1 Yr.	26%
1 Yr. or less	87.3%
Less than 6 Mo.	74%
Less than 1 Mo.	32%

The duration of symptoms varied from over 5 years to 1 day. Those lasting more than 1 year were 26 per cent; 1 year or less, 87.3 per cent; less than 6 months, 74 per cent; and less than 1 month, 32 per cent. The average duration approximated 8 months.

Symptom Area

	Cases	
Shoulder	66	(44%)
Shoulder and Neck	26	(17.3%)
Scapula	23	(15.3%)
Shoulder and Arm	22	(14.7%)
Arm and Hand	19	(12.7%)
Neck and Head	15	(7.3%)
Neck and Arm	2	(1.3%)
Bicipital Groove	2	(1.3%)
Chest	2	(1.3%)
Elbow	1	(0.67%)

The symptom area is shown. The shoulder was involved in 114, or 76 per cent, and, with the scapula, in 137, or 91.3 per cent, of the cases.

Fascia is a dense fibrous connective tissue that has the property of slight stretching and contracting particularly when a long-continued force is applied, and in this process stretches or constricts the structures that pass through or are attached to it. One important tissue passing through fascia is the spinal nerve that carries the afferent and the efferent impulses, which, when from an irritative source, are interpreted as pain.

The scapula is a suspended bone except for its articulation with the clavicle, which in itself is a bone suspended by soft tissues having the same or closely similar cord segmental innervations as do the muscles and fascia suspending the scapula. The muscles and the fascias that suspend the scapula all originate in the cervical or upper dorsal vertebrae, or are extension of the prevertebral and the postvertebral fascias that are penetrated by the spinal nerves. Any drag or sag exerted by the scapula in abnormal postural positions puts stress on these muscles and fascias, which in turn stimulate the penetrating nerves and initiate painful stimuli. Inman, Saunders and Abbott[1] state that motion occurs simultaneously in all joints of the region (shoulder girdle), each contributing its share. At the glenohumeral and the scapulothoracic articulations the ratio from almost the beginning to the termination of the arc is respectively 2 to 1. Steindler[5] states that abduction in the glenohumeral joint is associated from the beginning with scapular rotation.

The region of the scapula that has the greatest degree of motion, either translatory (that is, forward or back, up or down) or rotary, and also is supplied by the same cord segmental areas as the neck, the shoulder girdle and the upper extremity, is the area about the superior vertebral angle.

Tarsy[6] states that the spread of pain is

dependent on the intersegmental spread of noxious impulses within the cord These impulses are received by sensory neurons that are naked nerve terminals scattered throughout the deeper structures, subserving only pain. When the somatic lesion is persistent, these stimuli are relayed horizontally to the anterior horn cephalad and caudad for a number of segments, resulting in a pain spread beyond the bounds of the original segment involved. Injuries or diseases of the deeper structures, which in persistence or healing cause tissue tensions, give rise to chronic impulses to the cord that disturb the functional pattern of the internuncial pool of neurons. This cord mechanism is purely reflex and has not involved the higher centers, the thalamus or the cortex. Tarsy states that the concept of the vicious circle is a chain of interrelated forces, through the

TRIGGER POINT AREA

	Cases
Superior Vertebral Angle of the Scapula	90 (60.0%)
Spinous Process Base	40 (26.6%)
Scapula Verebral Border	20 (13.3%)

AMOUNT INJECTED
INTRACAINE B-DIETHYLAMINE ETHYL p-ETHOXY BENZOATE 2% IN OIL

Total Number of Injections: 254

1.0 cc. 14	2.5 cc. 122
2.0 cc. 60	3.0 cc. 58

	Cases
1 Injection	95 (63.4%)
Repeat Injections	55 (36 6%)
1 or 2 Injections	129 (86.1%)

In all these cases I used 1 per cent procaine in amounts of 2 to 10 cc., followed by Intracaine 2 per cent in oil in amounts of 1 to 3 cc. There were 254 injections in all. 2.5 cc. of Intracaine was used in 122 injections, or 49 per cent; 1 injection only was necessary in 95 cases, or 63 4 per cent; 1 or 2 injections were used in 129, or 86 1 per cent, of the cases.

POSTURAL TYPE

	Cases
Good	29 (19.0%)
Fair	48 (33.0%)
Fatigue and Poor	73 (48.0%)

The foregoing shows the general postural type of 150 patients. It is a somewhat arbitrary comparison and no doubt overlaps in a number of cases.

RESULTS

	Cases
Complete Relief	85 (56.6%)
Moderate Relief	50 (33.3%)
No Relief	15 (10%)

No Patient Was Made Worse
No General Reactions

With regard to the results, I wish to make clear here that in all cases in which postural abnormalities were noted, suitable physical therapy, both active and passive, was used persistently. In practically all cases not relieved by 1 injection (36.6%), physical therapy modalities were employed.

44 DIFFERENT OCCUPATIONS

Housewife	54	Broker	1
Rancher	14	Cameraman	1
Salesman	9	Chauffeur	1
Forewoman	4	Contractor	1
Office worker	4	Dietitian	1
Proprietor	4	Druggist	1
Retired person	4	Fisherman	1
Student	4	Grocer	1
Carpenter	3	Metal worker	1
Gardener	3	Miner	1
Maid	3	Packer	1
Physician	3	Plasterer	1
Truck driver	3	Postman	1
Janitor	2	Printer	1
Musician	2	Reporter	1
Nurse	2	Roustabout	1
Plumber	2	Sculptor	1
Secretary	2	Supervisor	1
Teacher	2	Union boss	1
Waitress	2	Unknown	1
Baker	1	Veteran	1
Bartender	1	Writer	1

This shows 44 different occupations. 93, or 69 per cent, in housewives, ranchers, salesmen, students, forewomen or office workers. There were even 2 musicians, 1 Union boss and 1 bartender.

establishment of which the primary disorder induces a second, which in turn aggravates the first. The trigger point or causal mechanism is established, and from this through cord segmental reflex phenomena new sources of pain are set up, possibly at some distance away. Treatment attack at the trigger point will interrupt the cord segmental flow of pain sensation to these reflex areas.

In a period of slightly less than 3 years in a private orthopaedic office practice I encountered 150 patients who had trigger points in the superior scapula area and exhibited pain patterns attributable to the scapulocostal syndrome.

SUMMARY

In summation may I say that I have attempted to present my experiences with 150 consecutive nonselected patients in a private orthopaedic practice over a period of 3 years. These patients all have had positive trigger points in the scapulovertebral area. No research other than reading was done. I believe that as regards this syndrome, the time is ripe for controlled experimentation and fundamental investigation in the field of histology, properties and pathology of fascia and the structures passing through and adjacent to it. I have postulated the etiology, the diagnosis and the classification of the scapulocostal syndrome, which might be called more accurately a scapulofascial syndrome, and presented 1 conservative injection method of treatment, along with a statistical analysis of the number and the site, the duration, the symptom area, the trigger point area, the amounts injected, the postural type, occupation and results. I believe that many of the unsolved problems now facing orthopaedic surgeons can be solved if they will recognize the scapulocostal syndrome, diagnose and treat it by whatever modality is most suitable for the individual patient.

REFERENCES

1. Inman, V. T., Saunders, J. B. DeC. M., and Abbott, L. C.: J. Bone & Joint Surg. *26*:1, 1944.
2. Michele, A. A., *et al.*: New York J. Med. *50*:1353, 1950.
3. Prentiss, H. J.: Regional Anatomy, pp. 2-4, Iowa City, Iowa Supply Co., 1922.
4. Russek, A. S.: J.A.M.A. *150*:25, 1952.
5. Steindler, A. L.: Kinesiology of the Human Body, Springfield, Ill., Thomas, 1955.
6. Tarsy, J. M.: Pain Syndromes and Their Treatment, pp. 23, 25, 27, 29, Springfield, Ill., Thomas, 1953.

Syndrome Scapulocostal

Summario in Interlingua

Le presente articulo describe le experientias clinic del autor con le syndrome scapulocostal in le curso de un periodo de tres annos, involvente 150 patientes consecutive in un private practica orthopedic. Iste syndrome—primo designate per su presente nomine per Michele in 1950—es un complexo dolente unilateral que involve le humero e le region scapulocostal con radiation del dolores verso le collo e le capite, le musculo deltoide, bracio e mano, e thorace.

Il pare exister duo typos: le typo atraumatic e le typo traumatic. Le prime ha un declaration insidiose inter le etates de 30 e 60 annos e occurre con tensiones postural o occupational sin historia de trauma. Le secunde ha un declaration plus acute e occurre post lesiones a plus juvene etates. Ambe typos ha acute punctos de precipitation al angulo supero-vertebral del scapula o al base de su processo spinose.

Es postulate que le etiologia debe esser vidite in un complexo resultante de contraction fascial, irritation nerval, e reflexo de nervo segmental. Omne le spinal truncos nervose in le nucha passa per le fascia profunde que es un parte del grande fascia profunde que coperi le integre corpore e es

continue per septos con omne le plus profunde structuras.

Le *punctos de precipitation in 86 pro* cento del casos se trova al angulo supero-vertebral del scapula. Es postulate que iste facto es explicabile si on considera que iste area del scapula indura le plus grande effortio suspensori super le fascia cervical e le musculos ubi es locate le nervos involvite in iste dolores de maniera que symptomas reflexe ab segmentos de chorda es referite a iste punctos.

Le scapula es un osso suspendite e articula con le altere duo ossos que forma le cinctura scapular. Le musculo deltoide es attachate a omne tres ossos sed multo plus al scapula que al alteres. Le musculo deltoide es involvite in 76 pro cento del configurationes dolente—o como region dolorose unic o in combination. Le sol histo commun que se applica a (e age super) omne le altere structuras profunde del collo—cinctura scapular, musculo, nervos spinal, fasces vascular, e septos intermuscular—es le fascia profunde.

Le total numero de patientes esseva 150. Cinquanta-duo pro cento esseva femininas, 48 pro cento masculos. Le latere dextere esseva involvite in 63,3 pro cento del casos e le latere sinistre in 36,7 pro cento. Le procentages de lateralitate pro le duo sexos esseva practicamente le mesmes. Le etate median esseva 45,09 annos—pro femininas 46,19 annos e pro masculos 44 annos. Le procentage de patientes con etates de inter *30 e 70 annos esseva 80,66. Le duration* median del presentia de symptomas ante le diagnose esseva octo menses. In 87,3 pro cento del casos iste duration habeva essite un anno o minus. Le area symptomatic includeva le humero in 76 pro cento del casos. Le area del puncto de precipitation esseva in le region del angulo supero-vertebral del scapula in 60 pro cento del casos.

Intracaina a 2 *pro cento in oleo in quan-*titates de 2,5 cm³ esseva usate in 49 pro cento del injectiones therapeutic. Un injection esseva requirite in 63,4 pro cento del casos; un o duo esseva requirite in 86,1 pro cento. Esseva representate 44 occupationes. Sexanta-nove pro cento del patientes esseva menageras, rancheros, commissos viagiator, studentes, e empleatos de bureau. Le resultatos obtenite monstrava alleviation complete in 56,6 pro cento del casos, moderate grados de alleviation in 33,3 pro cento, e nulle alleviation in 10 pro cento. Modos active e passive de therapia physic esseva usate conjunctemente.

Multes del non-resolvite problemas de dolores unilateral del nucha, del humero, e del extremitate superior es resolvibile si nos recognosce le syndrome scapulocostal e tracta lo per le modo therapeutic que es le melio applicabile al caso del patiente individual.

17

Problems Related to Prosthesis in Childhood

R. F. CHITTENDEN, M.D.*

INTRODUCTION

One meaning of the word *iconoclast* has to do with the breaking down of cherished ideals or long-revered beliefs. Because we have suffered for many years from the deterrent effect of beliefs about prosthesis in children (such remarks as "He is too young," or "He will outgrow it too soon," or "It isn't practical at that age") it seems now that it would be well for the iconoclasts to step forward and express themselves. There are many different ways in which this subject might be introduced. It is our basic philosophy that rehabilitation is not merely a word but a way of life for the incomplete individual, attained from within but aided from without by skillful guidance. We decry its use as a verb. We doubt that any therapist rehabilitates a patient. If you find this a difficult word to define, you will be in a position shared by many others. For the adult amputee I have presumed it to mean that rehabilitation was synonymous with a sense of security physically and mentally, attained through medical care, first-class limb fabrication, skillful physical therapy and good job counseling and training.

There is an established trend toward study of the special problems of the child amputee. This represents a very real contrast with the situation that has existed heretofore, in which the child generally was given attention relatively late, was put off about limbs for various excuses that are no longer valid, and certainly was *not* regarded with enthusiasm by the limb-maker. It is correct to assume that if the prosthetist did not advise limb-making for young children the physician was necessarily guided by this attitude and consequently deferred recommendation for fitting with the artificial member. The scope of the work with children does not indicate that an all-inclusive program has been established. It does mean that gratifying progress has been made, particularly in the centers already devoted to experimental limb prosthesis and rehabilitation as it relates to the very young individual.

It is not necessary to review the development of artificial limbs. Those who profess interest in such matters have already told us that when the warrior-knight of the Middle Ages survived the loss of a limb, his armorer constructed a completely articulated prosthesis for him, and examples of such work are still in existence. The idea of providing a substitute limb is therefore a very old one. Recent efforts to improve such devices have led more and more to the engineering laboratory. This does not mean that practical workers in the limb shop have not produced many interesting and useful devices applicable to the amputee, but it does mean that the trained engineer is now receiving deserved attention for his contributions, with benefit to the entire industry.

* Consultant to the Child Amputee Prosthesis Project, operated jointly by the Department of Engineering and the Department of Pediatrics of the School of Medicine, Medical Center, University of California, Los Angeles.

ETIOLOGIC FACTORS

Children need prosthetic aids because of congenital deformity, trauma and disease. Disease necessarily includes cancer, vascular and lymphatic anomalies that produce grotesquely enlarged limbs, infections, and paralytic processes of which poliomyelitis is the prototype. Trauma looms much larger after 6 years of age, and in situations where there is proximity to farm machinery. The very young children whom one encounters in a prosthesis clinic exemplify either congenitally arrested development or deformity or both. Before 2 years of age the factors of disease and trauma are numerically insignificant.

It is of interest to note that no age is too young for consideration by the rehabilitation center. We expect to see children selected for use of experimental prosthesis at a very early age, and even before that one will wish to prepare the parents for their role in the child's future and, if possible, to reassure them concerning his or her abilities and disabilities.

PSYCHOBIOLOGIC CONSIDERATIONS

There are several things that need to be kept in mind if one is being asked to approach the problem of the physically handicapped child. First of all, in the very young child, and particularly in some condition that has been present since birth, the problem is with the parents and not with the child, from the psychological standpoint. A little lad with an absence of part of one arm presents his parents with many misgivings and doubts, and they often blame themselves for his condition. They are greatly concerned about him and his future and they need much help in attaining assurance about him and reassurance for themselves.

From the standpoint of biology, very little is known about these odd arrests in limb development. They come very early in the growth of the unborn child, and in general it may be said that major derangements of structure that are going to resemble amputations of an extremity must have occurred before the first 3 months of pregnancy.

The parents are often in good health, with no history of physical abnormality in either family. They have 39 chances out of 40 of bearing a perfectly normal child in a subsequent pregnancy.

The child will function very well with one good hand. It is difficult to describe any real handicap, and inspection of his activities at play time will not reveal much impairment. The very young child, one may observe, is not much good at tying shoes or cutting up meat, whether he has one hand or two.

As has been said before, the child who has the need to be loved, the hope to be admired and the ability to respond to discipline may be regarded as a normal child. If the parents can meet these needs, they have provided a basically normal environment for the child. If they cannot, and are either rejecting the child or spoiling it by overindulgence, they will be guilty of contributing to whatever problem the child encounters, instead of lessening it.

The difficulties that one finds in dealing with the parents of the less-handicapped children are impressive. If a child lacks both arms completely, the acceptance of the fact of handicap cannot be avoided, and this of itself has some advantage at the intellectual level. The child who has underdeveloped fingers on one hand, by contrast, often acquires a habit of hiding the deformity, and the parents may indeed connive in this. Also, they may try to minimize the disability by saying that the hand is small rather than accept the fact that the child has almost no usable normal fingers on the extremity. By failure to be realistic and by underplaying the significance of the lesion, they may lay the foundation for certain psychological complexes.

It is not presumed to be necessary for the parents deliberately to draw attention to a physical defect. On the other hand, if it is noticed, there is no harm in acknowledging



FIG. 1. Congenital quadruple amputee, aged 5 years.



FIG. 2. Amputee with modern functional arm prostheses.

its existence, and it would be ridiculous to be evasive about it within the family or with close friends. We can regard parents as being secure and well adjusted when they treat the child well, provide a positive constructive program of activities in which the child participates, follow a considered disciplinary plan and acknowledge that a physical problem *does* exist.

The ultimate consummation of the constructive program and the most satisfactory expression of its components, so far as instruction is concerned, are embodied in the rehabilitation center.

UPPER EXTREMITY PROSTHESIS

THE CHILD FROM 6 TO 12

Many problems in upper extremity prosthesis have been recognized, but a number of these problems remain unsolved. The lack of appeal in the appearance of the functioning limb, when the wearer is dressed with short sleeves, is very well known. Except for purely cosmetic and almost completely functionless members, no solution has been offered. It may be all right to assume that when a person has lost both hands the urgency of the need of replacement is sufficiently great that this individual will not *protest cosmetic deficiencies in the artificial* limb. At the same time, if this person is appearing before the public the need for esthetic considerations is considerable, and the hook-type device continues to be forbidding, unless one is accustomed to it.

Factors contributing to rejection of prosthesis by the one-armed individual include poor function of the prosthesis, lack of

tactile sensation, poor appearance and discomfort to the wearer. The more extensive the amputation, the more difficult the replacement, and the factors of diminishing function become increasingly obvious as we approach complete absence of the upper extremity (Figs. 1 and 2).

The hook-type device works fairly well. It permits prehension of small objects better than the gloved hand or simulated fingers, is rugged enough to stand some abuse, and it provides a good training medium for the young amputees. Since the APRL hand is not generally available in any scaled-down size that would meet the needs of children under 12 years of age, the choice in terminal devices for the 6 year old necessarily lies between something resembling the Dorrance hook or some purely cosmetic nonfunctioning hand. The chief interest lies in *function* for the child amputee, although his ability to destroy the cosmetic hand must be considered.

The Infant Amputee

Exploration into the possibilities of prosthesis for infantile arm amputees has been attempted more and more frequently. One child, a bilateral congenital BE, with typical short below-elbow stumps, at 15 months of age, was referred to the Prosthesis Clinic in the Department of Pediatrics at the University of California, Los Angeles. The prescribing board concurred in the suggestion that he be fitted with typical BE limbs. This prosthetic equipment was a conventional BE with Dorrance hooks, but a "step up" type of elbow hinge was used on one arm and omitted from the other. Standard crossed-over shoulder harnessing was employed, and it became apparent that this lad, at 20 months of age, was completely capable of selecting which terminal device he wished to open, and of operating it correctly. It had been debated from experience with unilateral BE's at around 4 years of age that younger children would or would not operate shoulder harness effectively. It would be interesting for us to be able to say that this little boy, before 2 years of age, was quite skillful in the use of prosthesis, and dependent upon it for feeding himself, and that he found constant use for it. This is not true, and it obviously was in his way for many activities already learned without prosthetic aids. The fact that he rejected the limbs much of the time does not, in itself, justify any conclusions concerning the usefulness of prosthesis in young bilaterals.

Since that time we have seen several unilaterals at 15 to 18 months of age with upper extremity artificial limbs, using laminate constructions, standard harnessing and Dorrance terminal devices, in the "baby hook" category.

No doubt these children will be reported elsewhere and are mentioned in passing merely to emphasize the thought that the way to learn about arm prosthesis in infants is to practice it.

Problems of Acceptance

The factors concerned in rejection of arm prosthesis have been referred to already in this article. The author has under observation 2 girls aged 13; each was fitted with well-made limbs employing laminates, each was given both Dorrance hooks and APRL hands, both were exposed to identical periods of training in the use of the device. There was about 100% rejection in one child, and about 95% acceptance in the other. This would be very confusing if one did not make a careful study of these 2 children. Let us agree that initially each wanted an arm; that these were congenitals; that they were physically almost exact counterparts; and that they were living in the same city. There the resemblance ends. Our acceptor was able to list, before limb fabrication was undertaken, exactly what she thought the limb would do for her, and how she could use it. Our "rejector" is the product of a broken home, has the obvious disadvantage of a disinterested stepfather and a mother who tells her how badly the prosthesis looks.

Arm prosthesis for the school girl leaves much to be desired, if we are talking about appearance of the functioning device. Whether the patient is an AE or BE does not make too much difference because the elbow is rather revealing as to the mechanical nature of the arm substitute, and long sleeves become a near necessity for "dress up" occasions. Interchangeability of terminal devices may lead us in some instances to advise not less than 2 devices. One of these will be the hook: the other may be either the APRL hand or a nonfunctioning "purely" cosmetic one, such as the Realastic hand offered by Prosthetic Services of San Francisco. These "passive" hands can have some alteration made in the position of the fingers as regards flexion or extension but are completely lacking in voluntary control or prehension. This type of hand does represent a considerable advance in the technics of skin matching and undoubtedly is the least forbidding type of substitute, cosmetically speaking. It will receive favorable consideration by the adolescent amputees when they are before the public socially and when they are in situations where usefulness of a limb is not a consideration. We would prefer to teach these children the advantage of hook devices, try to elicit their co-operation in accepting them, and minimize the use of costly cosmetic devices of no functional value. However, there will be situations in which they will be used.

Those of us who advocate functional prosthesis are pleased with acceptance and annoyed by rejection. It remains fairly clear, however, that the girl patient with the short below-elbow stump will have extreme difficulty in masking her physical defect except when wearing long sleeves. We shall deprive her of tactile sensation when we apply prosthesis, and the degree of function obtained, although most helpful in some situations, will not enable her to play a stringed instrument or become a 2-handed touch typist and is not a true counterpart of the intact healthy hand. Therefore, acceptance of arm prosthesis cannot be taken for granted but must be sought after thoughtfully and with a well-co-ordinated program. Training and retraining with repeated exposure to other successful wearers may be important in achieving successful acceptance.

As regards trends in upper extremity prosthesis, I now believe that we shall accept very young children for experimental application of prosthesis; that totally armless infants will be fitted with elementary counterparts of prosthetic limbs in order to accustom them to restraint and to develop use of the shoulder girdle musculature; that small-sized scaled-down counterparts of the APRL hand will become available that will offer the child of 6 to 10 years cosmetically acceptable functioning prosthesis; that modified swim-fins will be developed for the special needs of the arm amputee, to assist in aquatic activities.

LOWER EXTREMITY PROSTHESIS FOR INFANTS AND CHILDREN

Lower extremity prosthesis in the not too distant past was quite unusual under five years of age and extremely rare before 3 years. Isolated instances of exception did not change the general impression that the very young children and infants were being allowed to go without limbs. In 1949, there was under observation a little lad—a congenital bilateral leg amputee — who was fitted with limbs at 10½ months of age and began walking at 11 months. His subsequent very satisfactory progress was reported in the *Crippled Child Magazine* (Feb., 1951) in an article by Dr. Donald Spiers, orthopaedist, and Gilbert Motis, of the Northrop limb research program. Considerable attention has been given to use of boots, pylons and various experimental limbs serving as prosthetic aids to infants. The commercial literature in the British Isles now frankly states that age 1 year is the time to set these children up with artificial limbs, an attitude with which, needless to say, we are in complete agreement. As was pointed

tactile sensation, poor appearance and discomfort to the wearer. The more extensive the amputation, the more difficult the replacement, and the factors of diminishing function become increasingly obvious as we approach complete absence of the upper extremity (Figs. 1 and 2).

The hook-type device works fairly well. It permits prehension of small objects better than the gloved hand or simulated fingers, is rugged enough to stand some abuse, and it provides a good training medium for the young amputees. Since the APRL hand is not generally available in any scaled-down size that would meet the needs of children under 12 years of age, the choice in terminal devices for the 6 year old necessarily lies between something resembling the Dorrance hook or some purely cosmetic nonfunctioning hand. The chief interest lies in *function for the child amputee, although his ability* to destroy the cosmetic hand must be considered.

The Infant Amputee

Exploration into the possibilities of prosthesis for infantile arm amputees has been attempted more and more frequently. One child, a bilateral congenital BE, with typical short below-elbow stumps, at 15 months of age, was referred to the Prosthesis Clinic in the Department of Pediatrics at the University of California, Los Angeles. The prescribing board concurred in the suggestion that he be fitted with typical BE limbs. This prosthetic equipment was a conventional BE with Dorrance hooks, but a "step up" type of elbow hinge was used on one arm and omitted from the other. Standard crossed-over shoulder harnessing was employed, and it became apparent that this lad, at 20 months of age, was completely capable of selecting which terminal device he wished to open, *and of operating it correctly. It had* been debated from experience with unilateral BE's at around 4 years of age that younger children would or would not operate shoulder harness effectively. It would be interesting for us to be able to say that this little boy, before 2 years of age, was quite skillful in the use of prosthesis, and dependent upon it for feeding himself, and that he found constant use for it. This is not true, and it obviously was in his way for many activities already learned without prosthetic aids. The fact that he rejected the limbs much of the time does not, in itself, justify any conclusions concerning the usefulness of prosthesis in young bilaterals

Since that time we have seen several unilaterals at 15 to 18 months of age with upper extremity artificial limbs, using laminate constructions, standard harnessing and Dorrance terminal devices, in the "baby hook" category.

No doubt these children will be reported elsewhere and are mentioned in passing merely to emphasize the thought that the *way to learn about arm prosthesis in infants* is to practice it.

Problems of Acceptance

The factors concerned in rejection of arm prosthesis have been referred to already in this article. The author has under observation 2 girls aged 13; each was fitted with well-made limbs employing laminates, each was given both Dorrance hooks and APRL hands, both were exposed to identical periods of training in the use of the device. There was about 100% rejection in one child, and about 95% acceptance in the other. This would be very confusing if one did not make a careful study of these 2 children. Let us agree that initially each wanted an arm; that these were congenitals; that they were physically almost exact counterparts; and that they were living in the same city. There the resemblance ends. Our acceptor was able to list, before limb fabrication was undertaken, *exactly what she thought the limb would do for her, and how she could use it.* Our "rejector" is the product of a broken home, has the obvious disadvantage of a disinterested stepfather and a mother who tells her how badly the prosthesis looks.

Arm prosthesis for the school girl leaves much to be desired, if we are talking about appearance of the functioning device. Whether the patient is an AE or BE does not make too much difference because the elbow is rather revealing as to the mechanical nature of the arm substitute, and long sleeves become a near necessity for "dress up" occasions. Interchangeability of terminal devices may lead us in some instances to advise not less than 2 devices. One of these will be the hook: the other may be either the APRL hand or a nonfunctioning "purely" cosmetic one, such as the Realastic hand offered by Prosthetic Services of San Francisco. These "passive" hands can have some alteration made in the position of the fingers as regards flexion or extension but are completely lacking in voluntary control or prehension. This type of hand does represent a considerable advance in the technics of skin matching and undoubtedly is the least forbidding type of substitute, cosmetically speaking. It will receive favorable consideration by the adolescent amputees when they are before the public socially and when they are in situations where usefulness of a limb is not a consideration. We would prefer to teach these children the advantage of hook devices, try to elicit their co-operation in accepting them, and minimize the use of costly cosmetic devices of no functional value. However, there will be situations in which they will be used

Those of us who advocate functional prosthesis are pleased with acceptance and annoyed by rejection. It remains fairly clear, however, that the girl patient with the short below-elbow stump will have extreme difficulty in masking her physical defect except when wearing long sleeves. We shall deprive her of tactile sensation when we apply prosthesis, and the degree of function obtained, although most helpful in some situations, will not enable her to play a stringed instrument or become a 2-handed touch typist and is not a true counterpart of the intact healthy hand. Therefore, acceptance of arm prosthesis cannot be taken for granted but must be sought after thoughtfully and with a well-co-ordinated program. Training and retraining with repeated exposure to other successful wearers may be important in achieving successful acceptance.

As regards trends in upper extremity prosthesis, I now believe that we shall accept very young children for experimental application of prosthesis; that totally armless infants will be fitted with elementary counterparts of prosthetic limbs in order to accustom them to restraint and to develop use of the shoulder girdle musculature; that small-sized scaled-down counterparts of the APRL hand will become available that will offer the child of 6 to 10 years cosmetically acceptable functioning prosthesis; that modified swim-fins will be developed for the special needs of the arm amputee, to assist in aquatic activities.

LOWER EXTREMITY PROSTHESIS FOR INFANTS AND CHILDREN

Lower extremity prosthesis in the not too distant past was quite unusual under five years of age and extremely rare before 3 years. Isolated instances of exception did not change the general impression that the very young children and infants were being allowed to go without limbs. In 1949, there was under observation a little lad—a congenital bilateral leg amputee — who was fitted with limbs at 10½ months of age and began walking at 11 months. His subsequent very satisfactory progress was reported in the *Crippled Child Magazine* (Feb., 1951) in an article by Dr. Donald Spiers, orthopaedist, and Gilbert Motis, of the Northrop limb research program. Considerable attention has been given to use of boots, pylons and various experimental limbs serving as prosthetic aids to infants. The commercial literature in the British Isles now frankly states that age 1 year is the time to set these children up with artificial limbs, an attitude with which, needless to say, we are in complete agreement. As was pointed

out, the advantages to the child, physically and mentally, and to the parents, outweigh any of the deterrents. The factors that worked against getting children fitted with limbs have included lack of precedent for this age group, clumsiness of conventional materials, apprehension concerning the effect of rapid growth on proper fitting, and a *disregard of the psychological importance of getting the child walking at an early age.* From a pediatric point of view our concern is with the whole child and with whatever might help to make him complete and functional in competition with other children of the same age. It is gratifying to see children walking alone with prosthetic aids that could not possibly have attained that goal at 14 months of age without them. Skilled therapists presumably could keep joints from developing contractures and build tone in muscles for children who are going without limbs—but actually one may obviate the need for such therapy and still conserve our physical therapists for more critical areas of endeavor by early use of prosthesis. The *parents,* too, will be very grateful.

Problems of Acceptance (Design)

The older girl amputees have been uttering objections to the prosthetic ankle and limb surface for many years. Many attempts have been made to construct an ankle that functions well and at the same time presents a good appearance under nylon hose and with pumps or flat shoes. Already these attempts have been rewarding, and we expect to see further improvement that will almost completely eliminate any notch or groove at the foot-ankle junction above the instep. Limb surfaces are now very often resilient, and improved coloring has been obtained for the shin section. As we suggested some time ago, in the 10 to 15 year age group, saddle shoes, ankle socks and rubber base paint to the shin piece surfaces give a very reassuring and pleasant appearance to the legs of girl amputees. At 15 years and older the wish to wear pumps and the need to escape from bobby-socks at times will make the need for an almost perfect ankle very apparent. The idea of formed toes with nails for the prosthesis seems to be an obvious further refinement for the prosthetic foot, and some prosthetists have obliged their girl customers with some ingenious woodcarving. It has occurred to the author that perhaps 8 or 10 stock sizes of the forefoot would cover the needs of 90 per cent of the patients and could be formed from tough plastics with skillful simulation of toes and nail structure.

In recent years the suction socket limb has gone from the novelty category into the classification of being the most frequently accepted prosthetic device for the above-knee child amputee. Included in successful user reports are children as young as 20 months of age, children with stumps 4 inches in length, and others, that have constituted pleasant surprises. The fact that some individuals do not wear this type of limb with success does not change the feeling about its general acceptance. Reports of suction socket developments for below-knee amputees continue to be made, and devices have been registered or patented covering that field. The author's personal experience with BK suction limbs in children does not place him in a position to make any comment upon them, but he would like to see such limbs used experimentally in the rehabilitation centers.

Children who completely lack a lower extremity may be able to make use of the device reported by Dr. Charles Hutter, with the saucer-type socket. It is apparent that this type of socket permits attachment of the limb in simpler fashion than the usual tilting table device and adapts itself to more natural sitting postures than the tilting table limb.

Very sincere attempts have been made to provide better kick mechanisms for the AK limb, as was reported by Catranis and others. The use of hydraulic plunger-piston accessories has been attempted both here

and abroad and undoubtedly deserves further attention. We are not aware of hydraulic-equipped limbs being used in children, but no doubt they would be applicable to the larger 12 year olds.

GROWTH FACTORS AND THE CHILD AMPUTEE

The factors that tend to lessen the adverse effects of growth in child amputees include the following: (1) The greatest growth occurs in the first year of life, and prosthesis is not needed before the close of that period. (2) Total anticipated linear growth is not a reliable guide to the problem, because growth in the head, the neck and the trunk does not contribute in any way to the difficulties. (3) In the affected portion, only the corresponding segments need be considered, because with few exceptions, remaining functioning epiphyses in the amputated extremity still operate to reduce the disproportion between affected and unaffected extremities. (4) It is possible to care for a little inequality in leg length by tilting the pelvis. (5) Some attention to the use of a "lift," either within or outside the shoe, can be used to correct a small disproportion between prosthesis and intact leg.

Upper extremity prosthesis is less of a problem concerning growth, although close supervision is desirable for all amputee children in order to ensure perfect fitting with prosthetic devices.

It is unfortunate that very few amputated limbs are end-bearing, and the distal termination of the stump must be relatively free in the limb socket. This means, for a thigh amputee, that a functioning capital femoral epiphysis produces growth in the stump only in terms of carrying it deeper into the limb socket of the artificial member and does nothing to help maintain alignment between the amputated and the nonamputated sides.

Experience shows that the difficulties of keeping an active 2 year old nicely fitted with prosthesis for leg amputation are considerable but not insurmountable, and the advantages to the child appear to outweigh other considerations by a clear margin.

AMPUTATION FOR DEFORMITY

One subject intimately related to the problem of limb prosthesis deserves more thoughtful attention than it has received. This is the policy of avoiding amputation or deliberately procrastinating definitive surgery in a patient presenting a limb that has an ultimately bad prognosis for appearance and function. When extremely rudimentary development is present in the femur, for example, one need not wait endlessly in order to forecast that the foot, if present on the affected side, is going to lie at about the level of the knee on the unaffected side. Reconstructive surgery, important though it may be, does not offer much to this patient. Walking may begin at 14 months of age with a modified long leg brace in which the foot and the deformed limb are laced into a "jacket" that takes most of the weight and may almost obviate the need for ischial seating in some instances. This type of device is purely functional and is cosmetically poor. Continuation of the use of this prosthetic substitute year after year because it is a good weight-bearing device and because it avoids any immediate need for surgery, and because it stimulates acetabular growth and hip joint formation, should not lead us to overlook the factors that make many of us wish to reject this type of apparatus in later childhood. The factor of psychic trauma is not a minor one. In some instances a poor decision or no decision may be most adverse to the well-being of the individual. This brace-prosthesis stamps one as crippled, particularly the girls, since it can be seen. If the foot lies at the level of the knee or immediately distal to it, the use of conventional artificial limb becomes practically impossible.

Recently the author has seen a girl of 16 years with a congenitally deformed right femur who has a good hip joint on the affected side. All that one can see in place of a right leg, when she is dressed for school,

out pubescence and still was sustaining the same increment 3 years after puberty, at a time when linear growth generally has "tapered off" in most female children. If we plot linear growth against age in years, then we obtain a straight line for the period of observation, from several years before pubescence until several years after pubescence, which is unexpected in terms of bodily growth. Is this therefore a counterpart of the overgrowth seen in osteomyelitis, and how often does it occur? We are well aware that in this same limb atrophy of the end of the bone has occurred and that the atrophy of hip joint on the affected side has resulted in an undersized femoral head. Thus, we may conclude that overactive epiphyses and underdeveloped bony structure may coexist, at least in some amputee individuals. (Fig. 4.)

The second subject is underdevelopment, which may be illustrated by the condition of the upper portion of the shaft and of the heads of the femora in a little boy; he was AK left and BK right from birth and walked on artificial limbs from the age of 11 months. At five years of age, in spite of constant use of the lower extremities in a very active little lad, we find that both upper femora are underdeveloped and that the comparison of his films with those of a child a full year

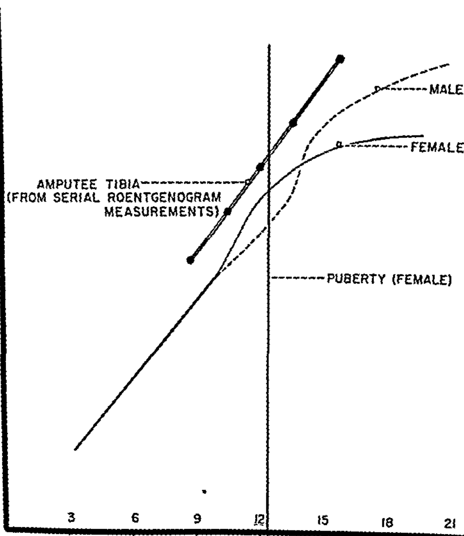


FIG. 4. Graph illustrating growth in fragment of tibia in girl leg amputee. Comparison with growth curves for linear body increment. One may note that in this patient there was no tapering off in growth pattern of the tibial growth increment in the period of observation. Menses in this patient began at 10 years and 10 months of age.



FIG 3. 17-year-old girl with brace for short femur.

is 2 metal bars terminating in a round pad about 3 inches in diameter. Cosmetically, this is on a par with Captain Ahab's peg leg, and when she is seated, since knee lock has been omitted, it must necessarily protrude in front of her, for everyone to see or to fall over. The orthopaedists are delighted that although she has worn this for 14 years, her hip development on the affected side was approximately identical with the unaffected one. (Fig. 3.) She has requested amputation, hoping that a standard suction socket prosthesis can be provided for her With the new arrangement all weight-bearing would be transferred to the tissues subjacent to the ischium We may then ask, of what great benefit is the well-developed femoral head and acetabulum since it is not to be used in weight-bearing with the new prosthetic set-up? A somewhat immature hip is not a "bad" hip from the standpoint of wearing an artificial limb and does not interfere with wearing a suction socket limb. The traumatic above-knee amputee who experienced amputation in early childhood has at puberty an underdeveloped femoral head and an immature femoral fragment. This has not proved to be any handicap, in the author's limited experience, in the fitting or the use of a suction socket limb, since the successful use of the prosthesis depends on range of motion and muscular development and not on the complete development of the bony architecture of the hip joint per se. This child would have had, we may presume, a much less difficult school experience if she had been amputated at 9 years of age, instead of 16. As has been said before, the child of 12 to 14 years is perhaps more anxious to present a good appearance, or a conforming appearance in relation to schoolmates, than any other age group we encounter.

One need not be led to the assumption that cosmetic and functional problems in congenital deformity cases are solved most frequently and easily by early amputation of the less useful portions of the affected limb. It is also true that no problem is solved by allowing a child to retain, year after year, an unsightly limb and an awkward and poorly fitting prosthetic device when the ultimate choice is still going to be amputation and fitting with a well-chosen artificial limb. Therefore, let us apply considered judgment to the individual case and remember what has been accomplished by judicious use of true surgical amputation.

PROBLEMS FOR FURTHER STUDY

Several matters related to growth and development in the child amputee deserve attention from the standpoint of research. A review of serial roentgenograms over a 10-year period may give us answers to matters that at present are entirely nebulous. One of these refers to overgrowth or hypertrophy in an amputation stump One series of roentgenograms on a girl amputee who became a BK through trauma at 14 months of age shows that linear growth in the tibial fragment proceeded at a uniform rate through-

out pubescence and still was sustaining the same increment 3 years after puberty, at a time when linear growth generally has "tapered off" in most female children. If we plot linear growth against age in years, then we obtain a straight line for the period of observation, from several years before pubescence until several years after pubescence, which is unexpected in terms of bodily growth. Is this therefore a counterpart of the overgrowth seen in osteomyelitis, and how often does it occur? We are well aware that in this same limb atrophy of the end of the bone has occurred and that the atrophy of hip joint on the affected side has resulted in an undersized femoral head. Thus, we may conclude that overactive epiphyses and underdeveloped bony structure may coexist, at least in some amputee individuals. (Fig. 4.)

The second subject is underdevelopment, which may be illustrated by the condition of the upper portion of the shaft and of the heads of the femora in a little boy; he was AK left and BK right from birth and walked on artificial limbs from the age of 11 months. At five years of age, in spite of constant use of the lower extremities in a very active little lad, we find that both upper femora are underdeveloped and that the comparison of his films with those of a child a full year

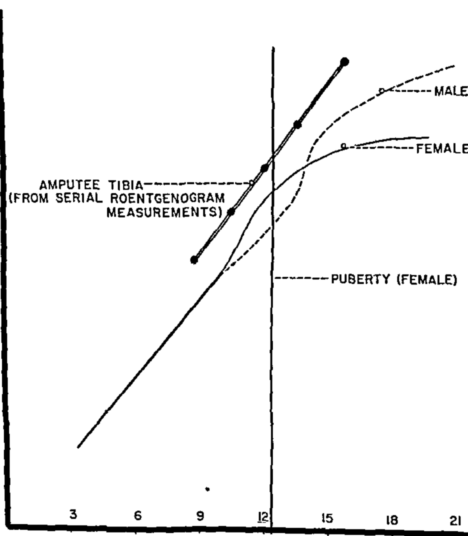


FIG. 4. Graph illustrating growth in fragment of tibia in girl leg amputee. Comparison with growth curves for linear body increment. One may note that in this patient there was no tapering off in growth pattern of the tibial growth increment in the period of observation. Menses in this patient began at 10 years and 10 months of age.

younger suggests that our patient is even less mature. This implies a lapse or retardation of about 2 years, which at 5 years of age is a very real disturbance. (Fig. 5.)

A 3rd topic in relation to growth and development concerns the need for study of the details of retardation pattern exhibited by unilateral congenital arm cases. The below-elbow cases not infrequently reveal underdevelopment in the supposedly unaffected segments, as for example in the upper arm of the affected limb. The question arises as to how much of this developmental arrest or retardation is from lack of use and how much is from an inherent lack of development due to congenital anomaly per se, that is to say, from genetic variation or intrinsic defect in limb structure on a congenital basis. It has seemed likely that comparison with teleoroentgenograms of postpolios that had acquired the disease severely in early infancy might give a lead toward answering this question, since we may presume that all the growth changes seen in the polio patient are due to acquired factors. Another interesting question is the nature of the anatomic detail of the defective segment, as in the BE stump. What muscles are present that correspond to the "normal" forearm? What are the muscles, the nerves and the vascular elements like in the area subjacent to the terminal portion of this limb? I have no presumption as to what the answer will be, but it does seem that studies could be directed to these subjects that might be rewarding to any interested physiologist, anatomist or orthopaedic surgeon.

Some considerable attention has been directed in the past several years to functioning braces for orthopaedically handicapped individuals. Because the patients are not amputees, this field lies outside the immediate scope of limb prosthesis but has a considerable overlap in technics and function objectives with those employed by the prosthetist serving the amputee. The totally flail upper extremity, like the total absence of an arm, or a forequarter amputation, remains a very challenging problem as regards restitution. Reports continue to appear in which prosthetic aids have been devised to impart function to totally flail upper extremities.

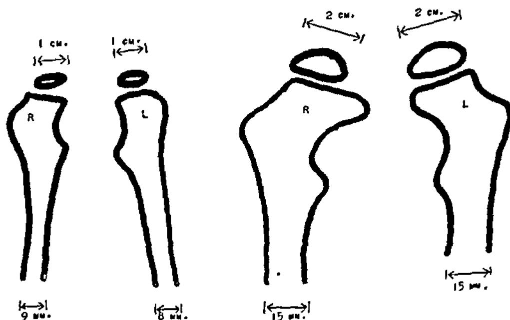


FIG. 5 Roentgenographic tracings of upper femora of amputee and intact child. *(Left)* Amputee (congenital) aged 4½ years, below knee and above knee. Walking with prostheses since 11 months of age. *(Right)* Intact male child aged 3½ years. No orthopaedic defect Walking since 12½ months of age.

Some of these devices are so nearly complete that one has the impression that the affected arm might as well have been left out of the device, and the prosthetic aid set up beside the patient to accomplish feeding or other acts by itself, under the paralyzed individual's control, but without incorporating his paralyzed extremity in its harness. More obviously successful use of prosthetic aids occurs where a hand has function but cannot be raised, or where the converse is true, in which the arm can be moved but the hand has no prehension. Devices applicable to these situations have been marketed, such as "Robin Aids" and others described in the literature, or as listed in the bibliography under Kent and Thompson.

It is very unfortunate that any child should come into this world with 2 or more extremities absent. Someone will say that it is doubly unfortunate that this child should survive with that degree of handicap. After a time this becomes very much aside from the point, since the child *is* surviving and deserves every possible attention in diminishing its disability and in helping it to adjust to a difficult physical situation.

DISCUSSION

There is no ineluctable need for the word *normal*. The author considers it a bad approach to address oneself to the parents of a child with a missing limb, saying, "In the normal child things are thus and so, and in your child this is going to be true" If the child in question fulfills all *other* criteria of normalcy, the missing limb should not be an excuse for setting him aside from his fellows. The ability of the individual to surmount difficulties and to rise above the restrictions supposedly imposed by physical limitations is no less remarkable than the failure of some of us to overcome very minor handicaps. Often it has been repeated that the bumblebee, from the standpoint of aerodynamics, should be unable to fly; its wings are too small, its body too blunt and heavy. I am thinking about a young man who lost both legs above the knee and eventually was able to derive considerable pleasure from ice-skating. He did not know that even unilateral thigh amputees are not supposed to be ice-skaters.

The amputee individuals constitute a numerically significant group. It can be estimated statistically that in a community with 9,000 postpolios needing rehabilitation there would be about 9,000 cerebral palsy cases in need of therapy care, and that this same community would have 14,000 amputees.

It was estimated that in 1955 the number of newborns would reach 5 million for the first time in our nation's history. This would lead us to expect that 1,760 children would be born in the United States with one or more limbs missing, to be added to our case load in that single year. The very youngest individuals are now receiving attention in the rehabilitation centers and observation in early infancy is no longer out of place. Emphasis on the use of laminates and plastic materials and better metals as well as the use of engineering methods in testing and limb fabrication, which was urged in our previous articles, has continued to prove its value and is gaining acceptance.

SUMMARY AND CONCLUSIONS

Problems in the amputee child have been reviewed from the point of view of etiologic and psychobiologic factors. Emphasis has been placed on upper extremity prosthesis in the older child and its role in the care of the infant patient. The acceptance of the artificial limb has been shown to be dependent upon many factors, which lead one to urge psychological preparation of the patient and his family, and the use of a well-guided training program.

The problems of growth and elective amputation are discussed. Children are generally adaptable and compensate for very bizarre handicaps. A strong competitive urge makes them good subjects for rehabilita-

tion and provides that necessary quality called "motivation." The young child, in contrast with some maimed adults, will strive very hard to keep up with or outdo his more fortunate playmates. Therefore, the work with children continues to be exceptionally rewarding.

BIBLIOGRAPHY

Abt, L.: Psychology of physical handicap, Orthop. & Prosth. Appl. J., p. 19, June, 1954.

Aitken, G., and Frantz, C.: Congenital amputation of the forearm, Ann. Surg. 141:519, 1955.

———: *The juvenile amputee, J. Bone & Joint* Surg. 35A:659, 1953.

———: Prosthesis for the juvenile amputee, Am. J. Dis. Child. 89:137, 1955.

Allredge, R.: Amputation and prostheses of upper extremities *in* Am. Acad. Orthop. Surgeons Instructional Course Lectures, vol. 10, Ann Arbor, Edwards, 1953.

Aylesworth, R.: Manual of Upper Extremity Prosthetics, Los Angeles, U.C.L.A., 1952.

Bluestone, S.: Rehabilitation of the handicapped child, Pediatrics 15:63, 1955.

Brooks, M., and Taylor, C.: First Ann. Report Child Amputee Prosthetics Project, Los *Angeles, Univ. California, Dec., 1955.*

Burnham, P.. Amputation of the lower extremity, *Ciba Symposium 6:143, 1954.*

Canty, T.: Cineplastic A-E Prosthesis: Internal Progress Report, Project Navy Prosth. Research Laboratory, NM 007 884.26, Nov., 1954.

———: *Construction Manual for Soft Socket* for Below Knee Artificial Limbs, Oakland, Calif., U. S. Naval Hospital.

———: Functional Ankle Joint *(Final Technical* Report), Vallejo, U.S. Naval Hospital, March, 1950.

———: Plastic Shins for Artificial Legs (Final Technical Report), Oakland, Calif., U.S Naval Hospital, Sept., 1954.

Chittenden, R.: Use of prosthesis for limb *defects in children, Am. J. Surg. 86:128,* 1953.

Dubinshak, J.: Children's prosthesis, Orthop. & Prosth. Appl. J., p. 27, Dec., 1952.

Frantz, C.: Prosthetic problems in juvenile amputees, Orthop. & Prosth. Appl. J., p. 13, 1952.

*Hutter, C.: Improved type of hip-disarticula*tion prostheses, J. Bone & Joint Surg. 35A: 745, 1953.

Kent, H.: Functional brace for the paralyzed hand, J. Bone & Joint Surg. *36A*:1082, *1954*

Klopsteg, P., and Wilson, P.: Human Limbs and Their Substitutes, New York, McGraw-Hill, 1954.

Rush, H., and Taylor, E.: Living With A Disability, New York, Blakiston Div., McGraw-Hill, 1953.

Scales, T.: Use of Durestos for orthopedic appliances, Plastics, July, 1948.

Steensma, J.: Training the juvenile amputee, *Orthop. & Prosth. Appl. J., p.* 18, Dec., 1952.

Thompson, T.: Forearm flexion device con*trolled by hip motor, Orthop. & Prosth. Appl.* J., p. 17, Sept., 1954.

Ware, E.: Parents of the orthopedically handicapped child (Mental Hygiene Series, No. 3), New York, Assoc. for the Aid of Crippled *Children, 1947.*

Problemas de Prosthetica in Juveniles

Summario in Interlingua

Iste articulo reporta le presente tendentias in le prosthetica pro infantes e juveniles Illo sublinea le problemas del amputato infantil e juvenil ab le puncto de vista del etiologia del deformitates, del importantia psychologic de iste conditiones pro le parentes, e del conditiones sub que adjutas prosthetic es utilisate. Attention es prestate al infante con amputation brachial, al factores que conditiona le rejection del prosthese, al necessitate del prompte application de adjutas del extremitates inferior, e al factores de crescentia e disveloppamento in amputatos. Mention es facite del amputation precoce como adjuta in obtener melior prostheses de gamba e del apparatos functional pro extremitates torpide in relation al campo del prosthetica.

18

Morphologic Variations of the Intercondylar Eminence of the Knee

BRUNO GIORGI, M.D.*

Roentgenologic investigation has opened a new field of research and study based on the ability to carry out observations on anomalies and variations of the skeletal system, a field which previously belonged almost exclusively to the anatomist.

It is not infrequent to perform a roentgenographic examination on individuals who are said to have established disturbances and to find a previous report of a skeletal anomaly which frequently has no relation to the presenting symptomatology.

We have had the opportunity to examine the roentgenograms of persons of both sexes and of varying ages who visited a specialist because of localized disturbances of the knee joint, and we have found varying degrees of variations in the morphologic development of the intercondylar eminence or the tibial spine.

As is known, the intercondylar eminence represents a small stubby fold located at the junction of the posterior third with the two anterior thirds of the tibial plate and extends into two elevations, the tubercles, one medial and one lateral. The intercondylar eminence assumes particular importance because of its anatomic relationship with the cruciate ligaments which have their insertion in it. These ligaments constitute the most important means of stabilization of the knee and of connection between the tibia and the femur.

The tibial spine takes origin from the secondary superior epiphyseal nucleus of the tibia which appears during the last 2 months of intra-uterine life. The elevation of the tibial spine appears at about the age of 2 years and gradually assumes the described quadrangular form as ossification progresses. Therefore, various degrees of morphologic variation can arise, related to anomalies in development. A review of the medical literature reveals that not a single author has investigated this subject specifically.

Only a German author, Bauer, maintains that there is a relationship between the form of the intercondylar eminence and the height of the articular rim with the specific purpose of securing an equilibrium at the joint. Indeed, he believes that all persons who manifest disturbances of the knee joint in whom one cannot find any definite disease may be assumed to have a change in the normal articular equilibrium which is symptomatically silent but radiologically well evident because of the altered relationship between the form of the tibial spine and the size of the joint space. Bauer had the opportunity to examine a significant number of roentgenograms and thus was able to study the variations in form of the intercondylar eminence. He subdivided these variations into 3 main groups.

*From the Istituto Ortopedico Toscano "Piero Palagi," Florence, Orthopedic Clinic of the University; Director, Prof. O. Scaglietti

tion and provides that necessary quality called "motivation." The young child, in contrast with some maimed adults, will strive very hard to keep up with or outdo his more fortunate playmates. Therefore, the work with children continues to be exceptionally rewarding.

BIBLIOGRAPHY

Abt, L.: Psychology of physical handicap, Orthop. & Prosth. Appl. J., p. 19, June, 1954.

Aitken, G., and Frantz, C.: Congenital amputation of the forearm, Ann. Surg. 141:519, 1955.

———: The juvenile amputee, J. Bone & Joint Surg. 35A:659, 1953.

———: Prosthesis for the juvenile amputee, Am. J. Dis. Child. 89:137, 1955.

Allredge, R.: Amputation and prostheses of upper extremities *in* Am. Acad. Orthop. Surgeons Instructional Course Lectures, vol. 10, Ann Arbor, Edwards, 1953.

Aylesworth, R.: Manual of Upper Extremity Prosthetics, Los Angeles, U.C.L.A., 1952.

Bluestone, S.: Rehabilitation of the handicapped child, Pediatrics 15:63, 1955.

Brooks, M., and Taylor, C.: First Ann. Report Child Amputee Prosthetics Project, Los Angeles, Univ. California, Dec., 1955.

Burnham, P.: Amputation of the lower extremity, Ciba Symposium 6:143, 1954.

Canty, T.: Cineplastic A-E Prosthesis: Internal Progress Report, Project Navy Prosth. Research Laboratory, NM 007 884.26, Nov., 1954.

———: Construction Manual for Soft Socket for Below Knee Artificial Limbs, Oakland, Calif., U. S. Naval Hospital.

———: Functional Ankle Joint (Final Technical Report), Vallejo, U.S. Naval Hospital, March, 1950.

———: Plastic Shins for Artificial Legs (Final Technical Report), Oakland, Calif., U.S. Naval Hospital, Sept., 1954.

Chittenden, R.: Use of prosthesis for limb defects in children, Am. J. Surg. 86:128, 1953.

Dubinshak, J.: Children's prosthesis, Orthop. & Prosth. Appl. J., p. 27, Dec., 1952.

Frantz, C.: Prosthetic problems in juvenile amputees, Orthop. & Prosth. Appl. J., p. 13, 1952.

Hutter, C.: Improved type of hip-disarticulation prostheses, J. Bone & Joint Surg. 35A: 745, 1953.

Kent, H.: Functional brace for the paralyzed hand, J. Bone & Joint Surg. 36A:1082, 1954.

Klopsteg, P., and Wilson, P.: Human Limbs and Their Substitutes, New York, McGraw-Hill, 1954.

Rush, H., and Taylor, E.: Living With A Disability, New York, Blakiston Div., McGraw-Hill, 1953.

Scales, T.: Use of Durestos for orthopedic appliances, Plastics, July, 1948.

Steensma, J.: Training the juvenile amputee, Orthop. & Prosth. Appl J., p. 18, Dec., 1952.

Thompson, T.: Forearm flexion device controlled by hip motor, Orthop. & Prosth. Appl J., p. 17, Sept., 1954.

Ware, E.: Parents of the orthopedically handicapped child (Mental Hygiene Series, No. 3), New York, Assoc. for the Aid of Crippled Children, 1947.

Problemas de Prosthetica in Juveniles

Summario in Interlingua

Iste articulo reporta le presente tendentias in le prosthetica pro infantes e juveniles. Illo sublinea le problemas del amputato infantil e juvenil ab le puncto de vista del etiologia del deformitates, del importantia psychologic de iste conditiones pro le parentes, e del conditiones sub que adjutas prosthetic es utilisate. Attention es prestate al infante con amputation brachial, al factores que conditiona le rejection del prosthese, al necessitate del prompte application de adjutas del extremitates inferior, e al factores de crescentia e disveloppamento in amputatos. Mention es facite del amputation precoce como adjuta in obtener melior prostheses de gamba e del apparatos functional pro extremitates torpide in relation al campo del prosthetica.

18

Morphologic Variations of the Intercondylar Eminence of the Knee

BRUNO GIORGI, M.D.*

Roentgenologic investigation has opened a new field of research and study based on the ability to carry out observations on anomalies and variations of the skeletal system, a field which previously belonged almost exclusively to the anatomist.

It is not infrequent to perform a roentgenographic examination on individuals who are said to have established disturbances and to find a previous report of a skeletal anomaly which frequently has no relation to the presenting symptomatology.

We have had the opportunity to examine the roentgenograms of persons of both sexes and of varying ages who visited a specialist because of localized disturbances of the knee joint, and we have found varying degrees of variations in the morphologic development of the intercondylar eminence or the tibial spine.

As is known, the intercondylar eminence represents a small stubby fold located at the junction of the posterior third with the two anterior thirds of the tibial plate and extends into two elevations, the tubercles, one medial and one lateral. The intercondylar eminence assumes particular importance because of its anatomic relationship with the cruciate ligaments which have their insertion in it. These ligaments constitute the most important means of stabilization of the knee and of connection between the tibia and the femur.

The tibial spine takes origin from the secondary superior epiphyseal nucleus of the tibia which appears during the last 2 months of intra-uterine life. The elevation of the tibial spine appears at about the age of 2 years and gradually assumes the described quadrangular form as ossification progresses. Therefore, various degrees of morphologic variation can arise, related to anomalies in development. A review of the medical literature reveals that not a single author has investigated this subject specifically.

Only a German author, Bauer, maintains that there is a relationship between the form of the intercondylar eminence and the height of the articular rim with the specific purpose of securing an equilibrium at the joint. Indeed, he believes that all persons who manifest disturbances of the knee joint in whom one cannot find any definite disease may be assumed to have a change in the normal articular equilibrium which is symptomatically silent but radiologically well evident because of the altered relationship between the form of the tibial spine and the size of the joint space. Bauer had the opportunity to examine a significant number of roentgenograms and thus was able to study the variations in form of the intercondylar eminence. He subdivided these variations into 3 main groups.

*From the Istituto Ortopedico Toscano "Piero Palagi," Florence, Orthopedic Clinic of the University; Director, Prof O. Scaglietti.



FIG. 1. Roentgenogram of normal knee of an adult. The intercondylar eminence appears normal in its anatomic profile with the tubercles standing out prominently, almost symmetrically.

The first variation which he believes is the most frequent one is characterized by a more prominent development of the medial intercondyloid tubercle as compared with the lateral tubercle.

In the second variation, the development of the 2 tubercles is about equal in height.

The third variation, which according to that author is the least common one, is characterized by a relatively greater height of the lateral tubercle. This variation, according to Bauer, is found especially in joints most markedly deformed.

Our studies of the variations in form of the tibial spine have been carried out by reviewing the roentgenologic case material of the Orthopedic Clinic of Florence.

We have examined 2,500 roentgenograms of the knee of individuals of both sexes, of young age, without signs of arthroses which in themselves could be responsible for structural changes. These patients had come for outpatient visits or had recovered from various joint disturbances. Our investigation was based exclusively on an examination of the radiologic findings, purposely ignoring the historical and clinical data.

We have taken into consideration only those roentgenograms which manifested variations in the form of the normal intercondylar eminence such as is represented in Figure 1.

In this way we have been able to establish sufficiently significant variations which we then were able to classify according to the following scheme (Fig. 2):

1. Anomalies characterized by a total or partial overgrowth
   - A. General hyperplasia
   - B. Hyperplasia of the medial intercondylar tubercle
   - C. Hyperplasia of the lateral intercondylar tubercle

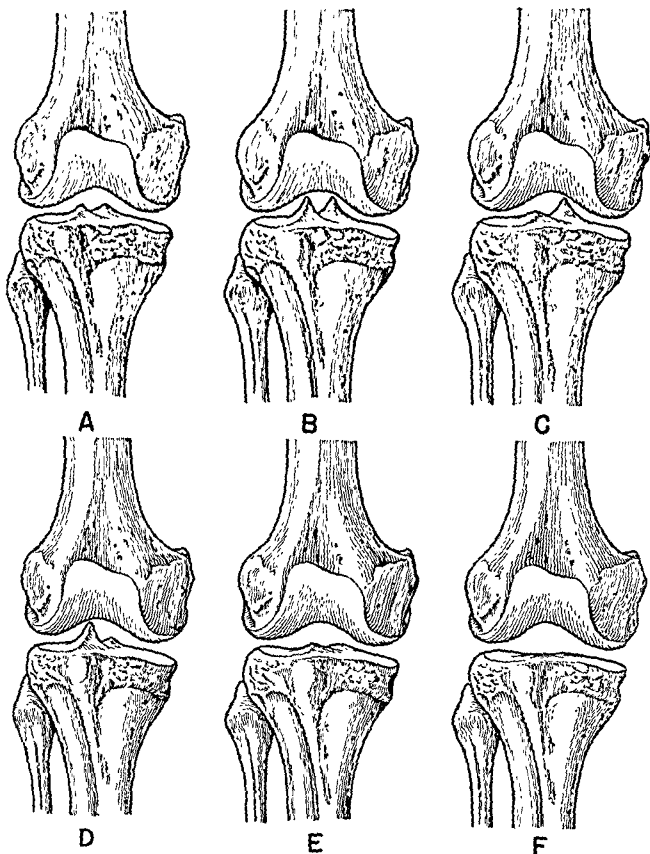


FIG. 2. Scheme of morphologic variations of the intercondylar eminence of the knee: (A) normal intercondylar eminence; (B) total hyperplasia of the intercondylar eminence; (C) hyperplasia of the medial intercondylar tubercle; (D) hyperplasia of the lateral intercondylar tubercle, (E) hypoplasia of the intercondylar eminence; (F) aplasia of the intercondylar eminence.



FIG. 3. April 4, 1954: Football (soccer) player, aged 24. Trauma to the left knee with locking of the knee in flexion. Physical examination: Left knee held in moderate flexion, swollen, tender to direct pressure on internal aspect of joint capsule. Extension not possible. No lateral movement. Roentgenographic examination revealed no osseous lesions. It was noted that the tibial spine was unusually prominent with both tubercles protruding well into the femoral intercondylar space. The diagnosis of a torn internal meniscus was made. The patient was operated on, the meniscus was removed, and there was complete recovery.

2. Anomalies characterized by defective development
   A. Hypoplasia
   B. Complete aplasia

Among the anomalies of excessive growth, the first variation is characterized by a greater development of the intercondylar mass with an abnormal and about equal prominence of the two intercondylar tubercles *protruding into the intercondylar space* of the femur (Fig. 3).

The second variation is characterized by a partial dysmorphism of the tibial spine manifested by abnormal development of the medial intercondylar tubercle which is excessive as compared with the lateral (Fig 4).

In the third variation one encounters an anomaly characterized by an abnormally prominent development of the lateral tubercle (Fig. 5).

On examining the anomalies characterized by defective development we have noted that in the first subgroup a flattening of the tibial spine is found with both tubercles definitely distinguishable in their contour but barely elevated (Fig. 6).

The second anomaly in this group *represented by a single case* was characterized by complete agenesis of the intercondylar elevation with a flattening of the superior surface of the tibial epiphysis without the slightest suggestion of bony elevation (Fig. 7).

Theoretically, this classification should be completed with cases of hypoplasia of one of the two tubercles. However, such a variation has not been encountered in our case ma-



FIG. 4. March 22, 1955: Female student, aged 17. Torsion injury to the left knee while skiing. Physical examination: The left knee appears to be normal in its contour. Moderate pain experienced on direct pressure on the region of insertion of the medial collateral ligament. Extension of the knee is not possible for about 10°. Flexion is possible to its full extent. No lateral movement. Roentgenographic examination failed to reveal any osseous lesions. Greater development in height of the medial than of the lateral intercondylar tubercle is noted. The diagnosis of a mild sprain of the knee was made. Treatment with rest alone resulted in complete recovery.

terial. From the various groups of anomalies we have selected the most characteristic cases of morphologic anomalies and have studied the clinical data of these cases.

It was found that of 5 individuals in whom roentgenologic examination revealed an anomaly, 4 had presented themselves for examination as the result of trauma which had occurred shortly before. Therefore, the finding of an anomaly was considered to be coincidental. This conclusion was also confirmed by the fact that once the responsible factor was eliminated all symptoms disappeared and complete recovery ensued. However, the case of complete aplasia of the tibial spine presented a different picture. In the patient in whom this anomaly was present the disturbances that originated in infancy without an apparent cause had become progressively severe. No traumatic factors had intervened which could have modified the course.

After these facts were established, the congenital absence of the tibial spine as a factor responsible for painful symptoms was taken into consideration. Indirect proof of this assertion was seen in the fact that the opposite knee presented a normal anatomic configuration and gave rise to no functional disturbances. From a clinical point of view the right knee showed a phenomenon of particular interest, namely, an abnormal mobility in the anteroposterior direction which was very marked ("drawer movement").



FIG. 5. June 3, 1953: Female farmer, aged 31. Contusion injury to the right knee. Physical examination: The right knee moderately swollen. No particular signs of pain. Flexion and extension movements are greatly limited. Roentgenographic examination failed to reveal the presence of any osseous lesions. Hyperplasia of the lateral intercondylar tubercle, as compared with the medial tubercle, was noted. The patient received a course of physiotherapy with regression of the symptoms described.

This finding, in addition to demonstrating that the feeling of instability complained of by the patient corresponded to an actual alteration of the mechanical condition of the knee, also suggests the possible relation of the absence of the tibial spine to a probable congenital alteration in the cruciate ligaments. Therefore, we hold that the cruciate ligaments in this case probably showed changes either as to their site of insertion or in their configuration. One could not explain otherwise the finding of marked ligamentous relaxation, provided that the ligaments were intact. Anatomic confirmation of such a hypothesis could have been furnished only by an exploratory arthrotomy which, however, was not performed because the indications were lacking.

Concerning Bauer's claim as to an existing reciprocal relationship between the height of the articular rim of the knee and morphologic variations of the tibial spine and the presence of a hyperplastic lateral intercondylar tubercle recognized by him in joints in which were noted severe signs of dysmorphism, we have not been able in our studies to find such relationships

From the standpoint of descriptive roentgenology we have found that, as a rule, the malformations are seen better in roentgenograms taken in the anteroposterior than in the lateral projection.

CONCLUSIONS

Concerning morphologic anomalies of the tibial spine we have found that numerous variations in form indeed exist which in the vast majority of cases are of small magnitude and of no significance and therefore may be considered as normal. However, in a small



FIG. 6. March 30, 1955: Female student, aged 18. Torsion injury to the left knee. Physical examination: Left knee swollen with signs of marked twisting of the joint. Pain felt on direct pressure on the insertion of the medial collateral ligament; exaggerated by bringing the knee into the valgus position. Roentgenographic examination failed to reveal the presence of osseous lesions. A morphologic variation of the tibial spine in the form of hypoplasia was noted. The opposite knee presented similar findings. The diagnosis of sprain of the knee was made, and a plaster cast was applied with resulting recovery.

number of cases, the variations take on characteristic appearances and can be classified as follows:

1. Hyperplasia of the tibial spine
2. Hyperplasia of only the medial tubercle
3. Hyperplasia of only the lateral tubercle
4. Hypoplasia of both tubercles
5. Complete aplasia of the tibial spine

On the basis of our clinical observations the anomalies characterized by hyperplasia and hypoplasia *do not cause alterations in* the mechanics of the knee joint productive of symptoms. Complete aplasia which, as we have stated, probably is associated with a morphologic and functional alteration of the cruciate ligaments is the cause of abnormal mechanical relationships which can lead to joint symptoms.

SUMMARY

The author has studied anomalies in morphology and development of the intercondylar eminence of the knee based on a large series of roentgenograms of the knee. He states that only in cases of complete aplasia *was it possible to demonstrate* alterations in the mechanics of the knee joint from a clinical standpoint.

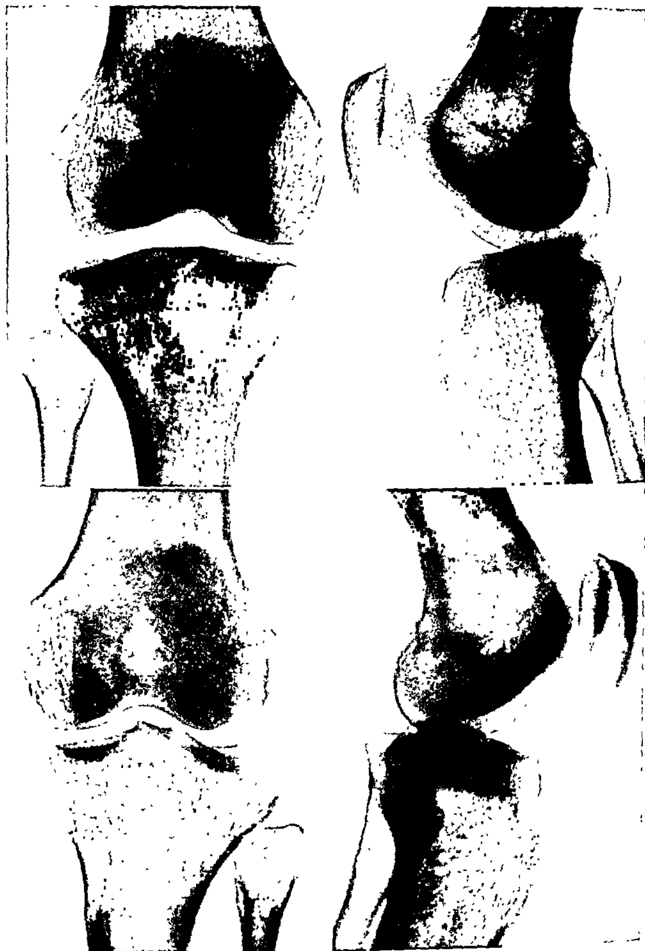


FIG. 7 (*Caption on facing page*)

Variationes Morphologic del Eminentia Intercondylar del Genu

Summario in Interlingua

Le autor ha studiate anormalitates del morphologia e del disveloppamento del eminentia intercondylar del genu in un grande numero de roentgenogrammas. Esseva constatate in iste eminentia, le spina del tibia, un multitude de variationes de forma, le quales, il es ver, ha minor dimensiones in le majoritate del casos. Alora illos es disproviste de signification e pote esser considerate como normal.

In un parve numero de casos, del altere latere, le variationes disveloppa formas characteristic que pote esser classificate sequentemente.

1. Hyperplasia del spina tibial.
2. Hyperplasia de solmente le tuberculo medial.
3. Hyperplasia de solmente le tuberculo lateral.
4. Hypoplasia de ambe tuberculos.
5. Aplasia complete del spina tibial.

Secundo nostre observationes clinic, le anormalitates hyper- e hypoplastic non affice le mechanica del articulation del genu de maniera a producer symptomas apparente. Quanto al aplasia complete, ilo es probabilemente associate con alterationes morphologic e functional del ligamentos cruciate e causa anormal relationes mechanic que pote resultar in symptomas articular a signification clinic.

FIG. 7. October 30, 1951: Female domestic, aged 32. The patient complained of having suffered, since infancy, a feeling of weakness and instability of the right leg with pains in the knee after prolonged activity. Physical examination: The right knee is normal in its anatomic profile. There are no signs of pain at the usual locations. On lateral movement both with the knee flexed and extended it is noted that an abnormal mobility is present as compared with the opposite leg. When the leg is forced into a position of flexion, there is noted at the condylar level a back-and-forth movement ("drawer movement") that is not present in the left leg. Roentgenographic examination of both knees revealed the following: (*top*) right knee—complete absence of the tibial spine; (*bottom*) left knee—a tibial spine normal in its anatomic outline.

19

Neurogenic Arthritis and the Problems of Arthrodesis of the Neurogenic Knee

L. W. WISEMAN, M.D.*

Because of its bizarre manifestations, neurogenic arthritis is a medical entity that has excited the interest of clinicians since its original description 100 years ago. The advent of newer and more effective modes of treatment of syphilis has so decreased the incidence of the condition that the average practitioner no longer is oriented to the problems of diagnosis and treatment. Sufficiently rare to be interesting, sufficiently common to be a problem, the destructive effect of the disease upon the involved joint and subsequent economic function of the victim are such that a review of the pathogenesis, the pathology and the treatment of the condition seems to be indicated.

The relationship between disease of the central nervous system and a destructive arthritis of the weight-bearing joints was first pointed out by Mitchell in 1831. Thirty-seven years later, Charcot first described the joint conditions that accompanied tabes dorsalis with sufficient clarity that this syndrome was given his name. In 1873 a case of poliomyelitis, complicated by neurogenic arthropathy, was described by Labrode. In 1875 Weir Mitchell described the condition in a case of myelitis, and in 1892 Sverdleff published a group of cases of syringomyelia with many examples of Charcot arthropathy.

Since its first description there has been considerable controversy concerning the pathogenesis of the neurogenic joint. Charcot stated that the syndrome was trophic in origin. Volkmann, and later Virchow, postulated that the changes characteristic of this disease arose on a mechanical basis as a result of the loss of the normal proprioceptive reflexes carried via the posterior roots to the spinal cord.

Numerous examples emphasize the importance of normal control by the central nervous system over peripheral structures. The deterioration of skin, bone and muscle in the paraplegic is well known; it does not seem to be unreasonable that a similar change in the articular structures would occur under the same environment. Definite trophic nuclei or centers have been described within the spinal cord near the lateral horn, and, in fatal cases of poliomyelitis with marked trophic involvement, extensive neuronolysis has been described in this area. The hypothesis has been that there originate from these nuclei peripheral vegetative fibers that are the same as, or are at least carried with, the main sympathetic trunks to the extremities. It is felt that their influence on the somatic structures is mediated by means of a control over the peripheral vascular system and, therefore, the nutritive supply to the periphery.[5,10]

Changes resembling neurogenic arthritis are found in paraplegic individuals who are not sufficiently ambulatory to provide mechanical trauma as the source of their disease; in addition, the joints of the upper

* San Gabriel, Calif

extremity frequently are involved in syringomyelia in which the factor of weight-bearing trauma is minimal. Furthermore, Charcot knees have been described in tabetics with proprioceptive reflexes still intact. The existence of atrophy of a single limb following congenital lesions of the parietal lobe is evidence of a specific trophic effect of the cerebrum on the growth and the development of the extremities. On the other hand, in most patients suffering from this disease there is definite impairment of their proprioceptive functions. Almost all so-called trophic ulcerations in syringomyeliacs were found by Grinker to have originated from trauma that would have been perceived and avoided by individuals with normal sensibilities. The continuation and the progression of the lesion then is favored by the inability of the patient to avoid repetition of the injury on already damaged tissues and by a crippled vasomotor system that is unable to provide the materials to overcome infection and produce repair. The higher incidence of these changes in the lower extremities, where the trauma of ambulation is almost incessant, likewise would appear to indicate a mechanical factor in its origin. It has been shown that section of the sciatic nerve in experimental animals never is productive of pathologic changes without superimposed trauma.[7,12]

In 1917, Eloesser carried out a series of experiments on cats that clarified the role of trauma in neurogenic arthritis. He showed that section of the posterior roots alone did not cause joint changes, but that section of the posterior roots plus traumatic insults to the analgesic joint resulted in a fulminating arthritis. It is significant that the majority of present-day investigations concerning the pathogenesis of this disorder have emphasized the importance of trauma. Probably it is best to consider the condition as an accelerated osteoarthritis precipitated by the application of trauma to a joint unprotected by proprioceptive and painful sensation.[6,8,12,13,14]

The pathology of this arthritis has been studied extensively. The first changes occur in the cartilage and underlying bone. There is an initial proliferative reaction of cartilage cells and matrix, followed by fibrillation, deterioration and formation of lacunae within the hyaline cartilage. Simultaneously, hyperplasia of the synovium and hypertrophy of the capsule develop as a result of recurrent synovial effusions. As a result, the capsule and surrounding ligaments are stretched and loosened so that subluxation, and even frank dislocations, can occur. Unlike ordinary varieties of osteoarthritis, the articular cartilage soon is invaded by a thick, destructive pannus originating from both the underlying bone and the synovial membrane. As the cartilage disappears, a revival of enchondral ossification occurs in an irregular, scattered fashion beneath the old articular cortex. Sclerosis and eburnation of the articulating bone ends take place, osteophyte production assumes grotesque proportions, and ectopic cartilage and bone appear throughout the synovial membrane. The degree of resumption of enchondral ossification is so great that by reason of simultaneous proliferation and resorption, an enormous incongruity of joint surfaces results with subsequent fragmentation of the articulating surfaces.[9,12,13]

As the disease progresses, it tends to resolve into either a hyperplastic or an atrophic form. The final picture in the hyperplastic form is one in which the disruption of the articular surfaces, ectopic bone formation, subluxations and dislocations have progressed to such grotesque proportions that they present an almost ludicrous appearance. In the atrophic form the joint debris is absorbed quickly, ectopic bone formation and calcification are minimal, and the articular surfaces appear to melt away, leaving a subluxated, unstable stub of a joint.

Neurogenic arthritis can occur following disruption of the normal proprioceptive protective reflex arc at any point from the joint to the cerebrum. It occurs most commonly in tabes dorsalis and syringomyelia. It has



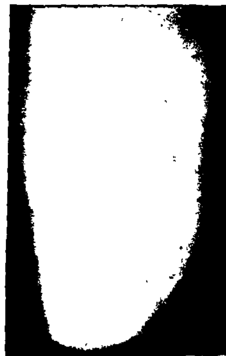


FIG. 1. (*Top, left*) Ill-advised and mechanically inefficient methods of arthrodesis in neurogenic arthritis are followed frequently by disaster. This patient was operated on for this severe neurogenic arthritis of the knee. (*Top, right*) In surgery, a crude approximation of the articular surfaces was performed. A long leg cast and an intramedullary nail were the only means of immobilization. (*Bottom*) With such haphazard immobilization and fixation, disaster was inevitable. 2½ months later, because of toxemia, gross purulent discharge and frank instability, a supracondylar amputation was performed.

been described following infectious, toxic, traumatic or neoplastic disruption of the peripheral nerves, nerve roots or the posteriolateral columns along their course in the spinal cord to the proprioceptive centers of the brain. Occasionally it is associated with pernicious anemia, paraplegia, diabetes, Pott's disease, cerebral tumors, poliomyelitis and chordotomy.[1,5,12]

This disease occurs usually in those between 35 and 65 years of age. The most common cause is tabes dorsalis, although there is a higher frequency of joint involvement in syringomyelia. The arthropathy is most common in Caucasians, despite the higher incidence of syphilis in the colored races; likewise, it is more common in men, although among women who develop tabes dorsalis the frequency and the severity of Charcot arthropathy are disproportionate. The usual time interval between the initial syphilitic infection and the onset of the disease in tabetics is 19 years.[8,13,14]

Usually the disease is associated with and preceded by signs and symptoms referable to the underlying pathologic process; however, in approximately one third of the cases the arthritis itself is the first subjective evidence *of the underlying neurologic disorder*. In almost every case a careful examination will disclose findings indicative of the presence of latent disease of the posterolateral columns. The vast majority of Charcot arthropathies are preceded in onset by the classic signs and symptoms of tabes dorsalis In one half of the cases there is serologic evidence of syphilis, although frequently serologic tests of the spinal fluid or blood alone may be negative.[8,13,14]

Neurogenic arthropathy may involve almost any articulation of the body, although it seems to show a predisposition for the weight-bearing joints, particularly the knee. Frequently it is polyarticular and bilateral in occurrence, although the sites of involvement are determined largely by the etiologic agent.

In 75 per cent of tabetics the disability develops in the lower extremity or the spine, while in over 80 per cent of syringomyelic arthropathy it occurs in the arm or the shoulder.[8,12,13,14]

Of course, the onset of the disease varies from joint to joint and from patient to patient. For instance, in the hip the initial symptom frequently is a pathologic fracture of the neck of the femur; in the spine it usually is crepitation with progressive disability and deformity; and in the knee, the ankle and the foot it generally is a sudden spontaneous swelling of the joint not preceded by great trauma or acute pain. In the knee the swelling may extend a considerable distance up and down the leg and may be associated with a dull, aching pain. The initial appearance of the joint may be such as to suggest a low grade inflammatory process. After a period of months, the effusion may clear only to recur continually as the process enters the chronic stage in which pain is minimal, abnormal mobility of the joint is present, and crepitation with valgus and varus deformities, subluxations and instability appear to be superimposed on the underlying bony destruction. As the disease progresses,



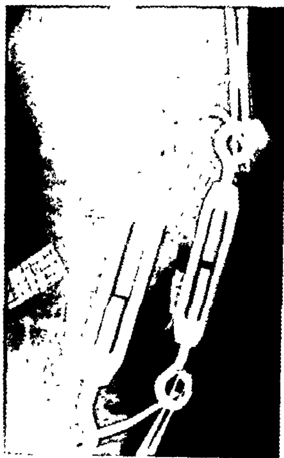




FIG. 2 (*Top*) A practical, stable and efficient compressive apparatus should be employed in compression arthrodesis. Steinmann pins of at least 3/32 in. in diameter should be employed and should not be bent beyond the limits of their elastic strength. This patient entered the hospital with this surprisingly mild neurogenic arthritis. (*Center*) At surgery, small 1/16 in. Steinmann pins were employed and were bent beyond the limits of their elastic strength, so that they could exert little actual compression. Crude hardware-store variety turnbuckles were used; they were attached to the pins only by hemostats. Only a long leg cast was used for postoperative immobilization. A postoperative roentgenogram showed what was to be expected—the usual anterior dislocation of the femur on the tibia. (*Bottom*) 1 year later, despite 2 attempted manipulations and a year in a hospital immobilized in a spica and one half, a definite nonunion of the arthrodesis still was present.

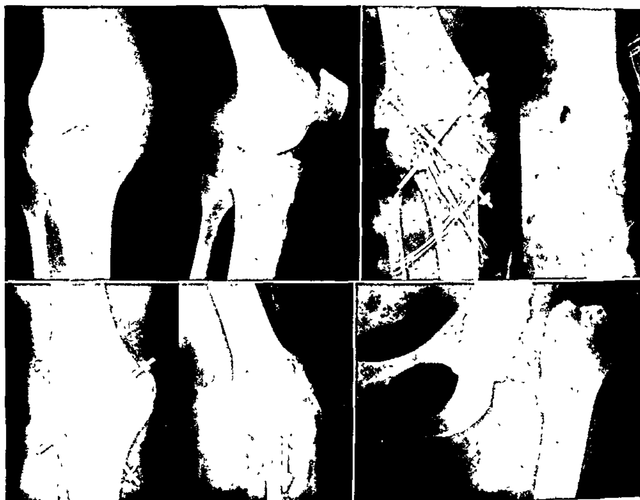


FIG. 3. (*Top, left*) Dislocation following arthrodesis has been found to occur in a definite and predictable fashion; that is, an initial extension of the tibia followed by an anterior dislocation of the femur. This patient was operated on for a mild neurogenic arthritis. A modified Galloway arthrodesis was performed, and the extremity was immobilized in a long leg cast.

(*Top, right*) 2 weeks after surgery a check roentgenogram showed extension of the tibia on the femur—an indication of imminent anterior dislocation of the femur. The arthrodesis was remanipulated and placed in a single spica cast.

(*Bottom, left*) 6 months later the arthrodesis was solid clinically and showed early union. Shortly afterward all external support was removed, and the patient began full, unprotected weight-bearing.

(*Bottom, right*) Neurogenic arthritis is a treacherous, unpredictable and systemic disease that often converts apparent success into dismal failure. Almost 1 year later, as the patient stepped from the curb, she suffered this pathologic fracture of the ipsilateral hip.

particularly in the knee and the foot, ambulation becomes more and more impossible, and gradual periarticular muscle atrophy develops. It is during this period that the victim becomes unusually susceptible to pathologic fractures of his long bones and periosseous soft tissue calcifications that add further to the deformity and the uselessness of the extremity.[1,9,13]

The therapy for this condition always has been a problem. Usually conservative treatment consists of antisyphilitic medication, bracing and muscle re-education in an attempt to protect the involved joint. More recently intra-articular hydrocortone has been administered without notable success and with the occasional complication of explosive pyarthrosis.



FIG. 4. (*Top*) A compression arthodesis, carried out carefully with accurate approximation of the articular surfaces, use of efficient turnbuckles and pins, and with postoperative immobilization of the arthrodesed knee in extension, will result in union of the arthrodesis in a high percentage of cases in a reasonable period of time. This patient had a severe neurogenic arthritis of the knee.

(*Center*) Arthrodesis of the knee was carried out with the knee in extension, large Steinmann pins bowed but not bent, efficient Charnley type turnbuckles and postoperative immobilization in a spica cast. (In later cases of this series, postoperative immobilization was supplemented by a Steinmann pin driven obliquely through the medial tibial condyle into the intramedullary cavity of the femur.)

(*Bottom*) 5 months later, after an uneventful postoperative course, a solid arthrodesis was present by both clinical and roentgenologic examination.

Direct surgical attack on neurogenic arthritis short of amputation always has been fraught with difficulties. The disastrous and frequently fatal effects of infection following surgery on these joints are well known. The avascular eburnated bone requires the best of surgical technic. The condition of the patient, his preoperative and postoperative care, and the long postoperative immobilization and rehabilitation require long and careful supervision by the surgeon and considerable forbearance and economic sacrifice on the part of the patient.

Arthrodesis is considered to be the surgical procedure of choice in this disease. Before surgery is performed the patient should be evaluated from the standpoint of his ability to survive the surgery and to withstand the long period of hospitalization. His general physical condition, with particular reference to the condition of the genito-urinary tract, the locomotive equilibrium status and the possibility of involvement of other joints, should be considered.

In the Los Angeles County Hospital a total of 8 arthrodeses have been performed for neurogenic arthritis of the knee since 1930; all were done during the last 10 years. In a similar period at the White Memorial Hospital in Los Angeles an additional 5 arthrodeses were performed for the same condition. This means a series of 13 patients with a total of 14 arthrodesing procedures; 1 patient was operated on twice. Eight were men, 5 were women.

The average age of patients at the time of surgery was 49; the youngest was 39; the oldest, 61. In 11 instances a diagnosis

tabes dorsalis was established conclusively by history, serology and general physical examination; in 2 cases a definite etiologic diagnosis of the basic neurologic disorder remained doubtful, but the history, the clinical findings and roentgenograms of the involved joint were typical of some type of neurogenic arthropathy. The average duration of symptoms prior to surgery was 3 years; the average time interval between the initial syphilitic infection and the onset of the arthropathy was 27 years.

In 5 cases there was evidence of multiple joint involvement, either at the time of surgery or as a subsequent complication. In 2 cases the additional joint involved was the hip; in 2 cases, the knee; and in 1 case, the mid-tarsal articulations. Ten of the 12 cases occurred in Caucasians and 2 in Negroes; of the 10 Caucasians, 5 were of Spanish extraction.

A review of the *Journal of Bone and Joint Surgery* and the *Annals of Surgery* reveals an additional 6 attempts at fusions of neurogenic knees from 1930 to 1954. If these cases are included, a group of 20 arthrodeses in 18 patients is produced, 2 patients having been operated on twice. A wide variety of arthrodesing procedures is represented: Albee, 1; Galloway, 1; Brittain, 1; arthrodesing procedure in which an intramedullary nail was used, 1; Soto-Hall, 1; Hibbs, 4; and compression-type arthrodesis as originated by Key and later modified by Charnley, 11—all done at Los Angeles County Hospital and White Memorial Hospital.

There were 6 nonunions in 20 arthrodeses. Three failures followed 11 compression arthrodeses, 1 followed 4 Hibbs procedures, 1 followed the use of an intermedullary nail and 1 occurred with the single Albee procedure. One compression arthrodesis at 7½ months still is incompletely united. Of the 3 failures with compression arthrodesis, 1 followed use of primitive and mechanically inefficient turnbuckles; the 2 remaining failures occurred in the same patient, and the 2nd nonunion probably can be regarded as an unsuccessful attempt to fuse a neoarthrosis. In addition, in the single Brittain procedure, distraction of the arthrodesis was noted at 3 weeks, and the subsequent salvage of the fusion can be attributed to the secondary application of turnbuckles to the cast and the use of the compression principle.

Four of the 18 patients had severe neurogenic arthritis at the time of surgery, and subsequently 2 of these developed nonunions. Of these 2 original failures, 1 was re-fused successfully. There is no significant difference in duration of symptoms prior to surgery between unions and nonunions. The average time of fusion in the 13 successful arthrodeses, calculated from the date of surgery to the time at which the surgeon considered the arthrodesis to be sufficiently solid to discard external support, was 8 months. The same period for the compression arthrodesis was 6½ months and for the Hibbs arthrodesis 5½ months; 1 compression arthrodesis was united at 3 months. The average duration of hospitalization was 14 days.

Postoperative complications developed in 21 instances. There were no fatalities as a result of surgery, although gross infection developed after 2 attempted arthrodeses, and 1 of them terminated in a supracondylar amputation. There were 18 complications among the 11 compression arthrodeses, including 7 pin-tract infections, 4 nonunions of the patella, 3 instances of distraction or gross dislocation of the arthrodesis, 1 late pathologic fracture of the hip following arthrodesis of an ipsilateral knee and 1 instance of difficult ambulation in a tabetic following prolonged bed rest. Seven of the 11 compression arthrodeses developed gross pin-tract infections; in only 1 instance was this sufficiently severe to necessitate discontinuation of the compression apparatus prior to the usual postoperative period of 2 to 3 months.

In evaluating the 11 compression arthrodeses, certain factors were found to be

of interest. The stability of the arthrodesis seems to be related to the accuracy with which the opposing surfaces are approximated and the extent to which the plane of the arthrodesis is kept at right angles to the line of compression. The use of a transverse incision and the application of a saw rather than an osteotome or a mallet appeared to facilitate these objectives. The maximum compression and fixation were obtained by driving the transverse Steinmann pins in close proximity and parallel to the arthrodesing surfaces.

The mechanism of distraction and dislocation of the 3 compression arthrodeses was by extension of the knee followed by anterior displacement of the femur. Whether the initial back knee is the result of the unopposed pull of the hamstrings and the gastrosoleus or simply the effect of gravity on the knee of a supine patient is uncertain. However, it does seem to be obvious that the maximum position of stability in compression arthrodesis is with the knee at 180°, and that flexion or extension beyond this point is productive of proportionate instability. In fact, it appears to be likely that once the initial angulation has occurred a continuation of compression will accelerate the deformity and complete the dislocation.

In each case distraction and dislocation were found to occur within the first 3 weeks. The use of a single Steinmann pin driven obliquely through the medial tibial condyle into the intramedullary cavity of the femur has proved to be a valuable supplement to the initial stability of the arthrodesis. The immobilization provided by a spica cast during this initial vulnerable period has been of great value and in 1 instance probably was the difference between success and failure. It is significant that no nonunions developed in those cases immobilized in a spica cast.

The use of small Steinmann pins and makeshift turnbuckles has been disastrous. The Steinmann pins should be at least 3/32 in. in diameter and should be connected by a mechanically efficient turnbuckle such as the Charnley device. Compression is exerted by shortening the turnbuckles periodically so as to maintain the Steinmann pins in a gentle bow; failure to maintain a bow or to tighten the turnbuckles to such an extent as to bend the pins has been found to decrease the compressive effect of the apparatus. Compression usually is maintained for 3 months, then the spica cast is removed, and the extremity is immobilized in a long leg cylinder cast or brace until the fusion is complete.

It is obvious that the small number of cases presented here makes the problem of drawing definite conclusions from this material extremely precarious. However, it is felt that certain tentative conclusions can be made even at this time:

1. Because of the wide variety of etiologic agents other than syphilis, neurogenic arthritis is, and will remain, sufficiently common so as to present problems in diagnosis and treatment.

2. Neurogenic arthritis probably is a fulminating osteoarthritis following the application of trauma to an ataxic joint.

3. Neurogenic knees can be fused successfully by a wide variety of methods; the disability attendant upon a neglected neurogenic knee is so severe that such fusion should be attempted.

4. A properly conducted compression arthrodesis supplemented by an obliquely driven Steinmann pin and application of a spica cast will arthrodese the neurogenic knee in a high percentage of cases within a reasonable period of time.

REFERENCES

1. Brailsford, J. F.: Radiology of Bones and Joints, ed. 5, pp. 701-704, Baltimore, Williams & Wilkins, 1953.
2. Campbell, W. C.: Operative Orthopedics, ed. 2, pp. 879-882, St. Louis, Mosby, 1949.
3. Charnley, J. C.: Positive pressure in arthrodesis of knee joint, J. Bone & Joint Surg. 30B:478-486, 1948.

tabes dorsalis was established conclusively by history, serology and general physical examination; in 2 cases a definite etiologic diagnosis of the basic neurologic disorder remained doubtful, but the history, the clinical findings and roentgenograms of the involved joint were typical of some type of neurogenic arthropathy. The average duration of symptoms prior to surgery was 3 years; the average time interval between the initial syphilitic infection and the onset of the arthropathy was 27 years.

In 5 cases there was evidence of multiple joint involvement, either at the time of surgery or as a subsequent complication. In 2 cases the additional joint involved was the hip; in 2 cases, the knee; and in 1 case, the mid-tarsal articulations. Ten of the 12 cases occurred in Caucasians and 2 in Negroes; of the 10 Caucasians, 5 were of Spanish extraction.

A review of the *Journal of Bone and Joint Surgery* and the *Annals of Surgery* reveals an additional 6 attempts at fusions of neurogenic knees from 1930 to 1954. If these cases are included, a group of 20 arthrodeses in 18 patients is produced, 2 patients having been operated on twice. A wide variety of arthrodesing procedures is represented: Albee, 1; Galloway, 1; Brittain, 1; arthrodesing procedure in which an intramedullary nail was used, 1; Soto-Hall, 1; Hibbs, 4; and compression-type arthrodesis as originated by Key and later modified by Charnley, 11—all done at Los Angeles County Hospital and White Memorial Hospital.

There were 6 nonunions in 20 arthrodeses. Three failures followed 11 compression arthrodeses, 1 followed 4 Hibbs procedures, 1 followed the use of an intermedullary nail and 1 occurred with the single Albee procedure One compression arthrodesis at 7½ months still is incompletely united. Of the 3 failures with compression arthrodesis, 1 followed use of primitive and mechanically inefficient turnbuckles; the 2 remaining failures occurred in the same patient, and the 2nd nonunion probably can be regarded as an unsuccessful attempt to fuse a neoarthrosis. In addition, in the single Brittain procedure, distraction of the arthrodesis was noted at 3 weeks, and the subsequent salvage of the fusion can be attributed to the secondary application of turnbuckles to the cast and the use of the compression principle.

Four of the 18 patients had severe neurogenic arthritis at the time of surgery, and subsequently 2 of these developed nonunions. Of these 2 original failures, 1 was re-fused successfully. There is no significant difference in duration of symptoms prior to surgery between unions and nonunions. The average time of fusion in the 13 successful arthrodeses, calculated from the date of surgery to the time at which the surgeon considered the arthrodesis to be sufficiently solid to discard external support, was 8 months. The same period for the compression arthrodesis was 6½ months and for the Hibbs arthrodesis 5½ months; 1 compression arthrodesis was united at 3 months. The average duration of hospitalization was 14 days.

Postoperative complications developed in 21 instances. There were no fatalities as a result of surgery, although gross infection developed after 2 attempted arthrodeses, and 1 of them terminated in a supracondylar amputation. There were 18 complications among the 11 compression arthrodeses, including 7 pin-tract infections, 4 nonunions of the patella, 3 instances of distraction or gross dislocation of the arthrodesis, 1 late pathologic fracture of the hip following arthrodesis of an ipsilateral knee and 1 instance of difficult ambulation in a tabetic following prolonged bed rest. Seven of the 11 compression arthrodeses developed gross pin-tract infections; in only 1 instance was this sufficiently severe to necessitate discontinuation of the compression apparatus prior to the usual postoperative period of 2 to 3 months.

In evaluating the 11 compression arthrodeses, certain factors were found to be

of interest. The stability of the arthrodesis seems to be related to the accuracy with which the opposing surfaces are approximated and the extent to which the plane of the arthrodesis is kept at right angles to the line of compression. The use of a transverse incision and the application of a saw rather than an osteotome or a mallet appeared to facilitate these objectives. The maximum compression and fixation were obtained by driving the transverse Steinmann pins in close proximity and parallel to the arthrodesing surfaces.

The mechanism of distraction and dislocation of the 3 compression arthrodeses was by extension of the knee followed by anterior displacement of the femur. Whether the initial back knee is the result of the unopposed pull of the hamstrings and the gastrosoleus or simply the effect of gravity on the knee of a supine patient is uncertain. However, it does seem to be obvious that the maximum position of stability in compression arthrodesis is with the knee at 180°, and that flexion or extension beyond this point is productive of proportionate instability. In fact, it appears to be likely that once the initial angulation has occurred a continuation of compression will accelerate the deformity and complete the dislocation.

In each case distraction and dislocation were found to occur within the first 3 weeks. The use of a single Steinmann pin driven obliquely through the medial tibial condyle into the intramedullary cavity of the femur has proved to be a valuable supplement to the initial stability of the arthrodesis. The immobilization provided by a spica cast during this initial vulnerable period has been of great value and in 1 instance probably was the difference between success and failure. It is significant that no nonunions developed in those cases immobilized in a spica cast.

The use of small Steinmann pins and makeshift turnbuckles has been disastrous. The Steinmann pins should be at least 3/32 in. in diameter and should be connected by a mechanically efficient turnbuckle such as the Charnley device. Compression is exerted by shortening the turnbuckles periodically so as to maintain the Steinmann pins in a gentle bow; failure to maintain a bow or to tighten the turnbuckles to such an extent as to bend the pins has been found to decrease the compressive effect of the apparatus. Compression usually is maintained for 3 months, then the spica cast is removed, and the extremity is immobilized in a long leg cylinder cast or brace until the fusion is complete.

It is obvious that the small number of cases presented here makes the problem of drawing definite conclusions from this material extremely precarious. However, it is felt that certain tentative conclusions can be made even at this time:

1. Because of the wide variety of etiologic agents other than syphilis, neurogenic arthritis is, and will remain, sufficiently common so as to present problems in diagnosis and treatment.

2. Neurogenic arthritis probably is a fulminating osteoarthritis following the application of trauma to an ataxic joint.

3. Neurogenic knees can be fused successfully by a wide variety of methods; the disability attendant upon a neglected neurogenic knee is so severe that such fusion should be attempted.

4. A properly conducted compression arthrodesis supplemented by an obliquely driven Steinmann pin and application of a spica cast will arthrodese the neurogenic knee in a high percentage of cases within a reasonable period of time.

REFERENCES

1. *Brailsford, J. F.:* Radiology of Bones and Joints, ed. 5, pp. 701-704, Baltimore, Williams & Wilkins, 1953.
2. Campbell, W. C.: Operative Orthopedics, ed. 2, pp. 879-882, St. Louis, Mosby, 1949.
3. Charnley, J. C.: Positive pressure in arthrodesis of knee joint, J. Bone & Joint Surg. 30B:478-486, 1948.

4. Cleveland, M., and Smith, W.: Fusion of the knee joint in cases of Charcot's disease, J. Bone & Joint Surg. 13:849, 1931.
5. Delano, P. J.: Pathogenesis of the Charcot joint, Am. J. Roentgenol. 56: 189-200, 1946.
6. Eloesser, L.: On the nature of neuropathic affections of the joints, Ann. Surg. 66:201-207, 1917.
7. Grinker, R. R.: Neurology, ed. 2, Springfield, Ill., Thomas, 1937.
8. Key, J. A.: Clinical observations on tabetic arthropathy, Am. J. Syph. 16: 429-446, 1932.
9. Luck, J. V.: Bone and Joint Disease, ed. 1, p. 243, Springfield, Ill., Thomas, 1950.
10. Nielson, J. M. A.: Textbook of Clinical Neurology, New York, Hoeber, 1941.
11. Soto-Hall, R.: Fusion in Charcot joint of the knee, Ann. Surg. 108:124-126, 1936.
12. Soto-Hall, R., and Haldemann, K. O: Diagnosis of neuropathic joint disease, J.A.M.A. 114:2076-2078, 1940
13. Steindler, A.: The tabetic arthropathies, J.A.M.A. 96:250-256, 1931.
14. Wile, U. J., and Butler, M. G.: A critical study of Charcot arthropathy, J.A.M.A. 94:1053-1055, 1930.

Arthritis Neurogene e le Problemas de Arthrodese del Genu Neurogene

Summario in Interlingua

In despecto del reducite incidentia de syphilis, arthritis neurogene remane sufficientemente commun pro esser de interesse pro le chirurgo orthopedic. Iste lesion occurre in association con non importa qual morbo que interrumpe le protective arco de reflexo proprioceptive a ulle puncto al longo del peripheria del medulla spinal. Le lesion se incontra ancora le plus frequentemente in association con tabes dorsal, sed illo es etiam associate con numerose casos de anemia perniciose, diabete, e syringomyelia. Su melior description esseva forsan su characterisation como osteoarthritis fulminante occurrente post le application de trauma a un articulation ataxic.

Arthrodese es considerate como le tractamento de election pro patientes con arthritis neurogene. Tamen, il es difficile obtener resultatos successose in tal patientes. Le rationes pro iste difficultate include (1) le tendentia del morbo de manifestar se, post su tractamento chirurgic, in altere articulationes, (2) le aberrante e anormal physiologia associate con le subjacente disordine neurologic, (3) le presentia de sclerotic osso inactive que es susceptibile al infection, e (4) le prolongate immobilisation e rehabilitation postoperative que es necessari.

Isto es un reporto de un serie de deceocto patientes in qui vinti arthrodeses esseva executate pro arthritis neurogene del genu. In dece-quatro casos le arthrodese resultava in un fusion successose.

Arthrodese a compression es apparentemente le plus frequente methodo therapeutic in iste morbo. Quando iste arthrodese es ben executate e supplementate, illo resulta in fusion intra un periodo non excessive in un alte procentage del casos.

20

Deep Thrombophlebitis, A Cause of Acute Synovitis of the Hip Joint*

E. Dyer Davis, M.D.†

I shall present 2 cases of clinical thrombosis of the deep veins for the specific purpose of establishing an etiology for syndromes consisting of pain, muscle spasm and limitation of motion about the hip joint.

Case 1. Negro, aged 23, past history noncontributory. The patient entered the hospital on January 9, 1956. His only complaint was excruciating pain in the left hip. Physical examination revealed extreme tenderness of the left hip at the intertrochanteric fossa posteriorly. Any active or passive movement of the leg precipitated agonizing pain with severe spasm of all muscle groups of the thigh. He was unable to bear weight. No tenderness was noted over the great vessels anteriorly. Significant laboratory findings were: elevated white count to 10,500; blood urea nitrogen 48 mg. per cent and 3 plus protein in the urine.

There was little change with conservative treatment of bed rest, sedation and Buck's extension. Four days later the left thigh appeared to be grossly swollen. Compression dressings were applied, and anticoagulant therapy was started. Despite therapy, the thigh continued to enlarge, and, 2 weeks after admission, by actual measurement it was 4 in. larger in circumference than the right.

At this time the hip pain continued to be severe, and persistent 3 plus protein in the urine and a reversal of the albumin-globulin ratio were noted. Not until February 2, 4 weeks after admission to hospital, was there any diminution of symptoms about the hip joint. Three days after the beginning of improvement, the patient's hip pain was gone. However, his clinical picture was one of a typical post-thrombophlebitic leg with swelling and induration.

It is assumed that the acute symptoms referable to the hip joint were the result of venous engorgement from this process or an extension of the actual inflammatory process into the vascular system of the hip joint.

Case 2. Negress, aged 18. This patient developed pain in the right hip 2 weeks after normal vaginal delivery. On admission to hospital she was unable to stand and shrieked with pain on attempted manipulation of the leg or on palpation of the intertrochanteric fossa posteriorly. The remainder of the physical examination was normal except for a pulse rate of 120 per minute. A significant laboratory finding was an elevated white count of 13,600.

Four days after admission the right thigh appeared to be swollen grossly, and by measurement was 2 in. greater in circumference than the left. Treatment was conservative with bed rest, elevation, Ace bandage and Buck's extension. By the end of 2 weeks the patient was asymptomatic, although swelling of the leg persisted. She was discharged 21 days after admission, and on follow-up 4 weeks later the thighs were of equal circumference.

Thrombosis is the result of an upset in a delicate chemical and physical balance of circulating elements and vascular endothelium. Some of the known predisposing fac-

* Read at meeting of the Philadelphia Orthopaedic Society April 6, 1956.

† Resident, Philadelphia General Hospital.



FIG. 1. Postero-anterior view of the pelvis (Case 1) shows no abnormalities of either hip joint. Four subsequent roentgenograms at weekly intervals failed to show any change.



FIG. 3. Venogram done through a femoral puncture 2 weeks after admission to hospital (Case 1). Observe the presence of dye in the left femoral and iliac veins with failure to visualize dye passing into the inferior vena cava, indicating a block of the deep venous system on the left. Observe the hypertrophied anastomotic vessels crossing the anterior portion of the pelvis and entering the deep venous system of the opposite side through the hypogastric and the iliac veins.

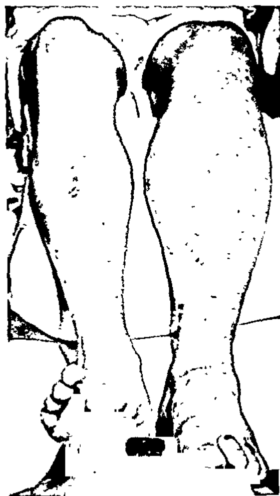


FIG 2 Lower extremities 2 weeks after admission to hospital (Case 1). Note the difference in circumference of the 2 legs—4 in. in the thighs, 2 in. in the calves.

tors of thrombus per se are injury to the intima, slowing of the blood stream and change in chemical composition of the blood.

The most usual location of thrombus formation in the lower extremities is in the plantar veins, the veins of the calf muscles, the veins of the adductor muscles and the visceral pelvic veins, especially in obstetric patients.

In neither Case 1 nor Case 2 was there objective or subjective evidence of thrombosis anywhere except in the *high femoral* or the iliac veins after the hip syndromes were fully developed. The etiology of Case

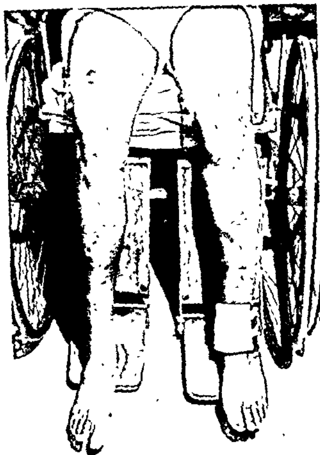


FIG. 4. Lower extremities 6 weeks after admission to hospital (Case 1). Patient had had treatment of compression dressings, elevation and anticoagulant therapy.



FIG. 5. Postero-anterior view of the pelvis of Case 2 showed no abnormalities at the time of admission to hospital. Follow-up roentgen examinations also were normal.

2 is rather clear cut, since the onset was 2 weeks after delivery. No such evidence is observed in Case 1. However, in both cases the onset and the character of the hip syndromes are strikingly similar.

J. Benassy and J. Lamare,[2] of Paris, have injected a series of normal living femurs through the greater trochanter with radiopaque dye to establish a norm for venous drainage of the hip joints. In the normal hip it was found that the drainage occurred principally through the external iliac vein, the obturator system and the superior and the inferior gluteal system. In cases of osteoarthritis of the hip, neither the femoral nor the iliac veins were visualized. The collaterals took nefarious routes through complex anastomoses into the hypogastric systems of the opposite side, much in the same manner as is illustrated in Figure 3. These authors have stated that, besides the ischemic lesions of arterial origin proved by Trueta, thrombosis of the circumflex or the obturator veins may play a role in the etiology of certain cases of arthritis affecting the hip joint.

An interesting side light is the renal picture accompanying the syndrome in Case 1, which is highly suggestive of renal vein thrombosis. Both in animal experiments[4,7,8] and in patients proved at postmortem to have had renal vein thrombosis, there was evidence of great protein loss in the urine with a reversal of the albumin-globulin ratio.[3,5,6] The patient in Case 1 had changes in the plasma and the urine characteristic of a nephrotic syndrome and could be distinguished from it only by the absence of a suggestive clinical history and by its ephemeral nature. This interference with the renal venous return presumably developed slowly, since rapid occlusion usually causes hemorrhagic infarction of the kidney.[1]

To my knowledge, the clinical picture of acute synovitis of the hip joint as the first manifestation of thrombophlebitis of the deep veins has not been reported in the literature. On admission to hospital, and for several days thereafter, Case 1 had the primary symptoms and the clinical findings of an acute process confined to the left hip

joint. He complained of pain in the hip, excruciating pain on palpation, agonizing pain with any movement of the joint, either actively or passively. There was severe spasm of all muscle groups of the thigh and never a history of calf or thigh pain. Case 2 showed the identical symptoms in regard to the hip pain. Subsequently swollen thighs and calves developed in both cases from 4 to 6 days after admission. Both hips were aspirated before antibiotics were administered, and the cultures were reported as no growth. In both cases blood cultures failed to reveal bacteremia. Viral antibody studies did not show a significant rise in titer.

The lack of any positive findings in the above studies seemed to indicate that the syndrome described was of vascular origin. Thus, a definite etiologic factor is established in those cases in which the syndrome resembles acute transient synovitis of the hip.

REFERENCES

1. Bell, E. T.: Renal Diseases, ed. 2, London, Kimpton, 1950.
2. Benassy, J., and Lamare, J.: Rôle de la thrombose veineuse dans la pathogénie de la coxarthrose, Rev. rhum. 22:59-67, 1955.
3. Blainey, J. D., *et al.:* Nephrotic syndrome associated with thrombosis of the renal veins, Lancet 2:1208-1211, 1954.
4. Blake, W. D.: Effect of increased renal venous pressure on renal function, Am J. Physiol. 157:1-12, 1949.
5. Derow, H. A., *et al.:* Chronic progressive occlusion of the inferior vena cava and the renal and portal veins, Arch. Int. Med. 63:627-647, 1939.
6. Osler, W.,- and McCrae, T.: Modern Medicine, Its Therapy and Practice, ed. 2, p. 789, Philadelphia, Lea and Febiger, 1914.
7. Robinson: Med. Chir. Tr. 26:51, 1843.
8. Rountree, Fitz and Geraghty: The effects of experimental chronic passive congestion on renal function, Arch. Int. Med. 11:121-147, 1913.

Thrombophlebitis Profunde, un Causa de Acute Synovitis del Articulation Coxo-Femoral

Summario in Interlingua

Es reportate le casos de duo patientes qui exhibiva al tempore del prime presentation le classic signos e symptomas de acute synovitis del articulation coxo-femoral. Le prime patiente esseva un masculo de 23 annos de etate, le altere un feminina de 18 annos, duo septimanas post parturition.

Culturas del fluido synovial, culturas de sanguine, e studios de anticorpores viral non produceva resultatos positive. Subsequentemente ambe patientes disveloppava thrombophlebitis del profunde systema venose in le extremitate al latere del coxa afficite. Un constatation incidental esseva thrombosis del venas renal in le masculo de 23 annos.

Le absentia de resultatos positive in le supra-mentionate studios pareva indicar que il se tractava de un syndrome de origine vascular. Assi se establi un definite factor etiologic in iste casos in que le syndrome es un acute synovitis transiente del coxa.

21

Pathways Tracked by Dorsolumbar Tuberculous Abscesses

ROBERT L. SAMILSON, M.D.*

Since the advent of potent antituberculous drugs, the diagnosis and the subsequent treatment of paravertebral tuberculous abscesses has been altered greatly. The frequency of indolent sinus tracts has all but disappeared. At the New York Orthopaedic Hospital, a number of tuberculous abscesses originating in the dorsolumbar spine have been studied recently by the direct injection of radiopaque dye substances. A graphic demonstration of the anatomic pathways tracked by these abscesses has resulted.

The incidence of "cold" abscesses associated with vertebral tuberculosis varies from 20 per cent[2] to 63 per cent[3]. Apparently, this sequela is more frequent in lower dorsal and lumbar disease than in other spinal segments. In a recently reported series from this institution, Hallock and Jones[1] found that 91 per cent of spinal tuberculosis originated in the lower dorsal, the dorsolumbar, or the lumbar segments.

ANATOMIC PATHWAYS

Figure 1 depicts the 3 common pathways tracked by abscesses from the lower dorsal or lumbar spine. Perhaps the most frequent route is within the sheath of the psoas muscle, to present in the iliac fossa, or to gravitate downward below the inguinal ligament, usually on the adductor aspect of the thigh. The abscess may penetrate either the anterior longitudinal ligament of the spine or the ventral psoas sheath to gain access to the retroperitoneal space. From there, several routes may be taken. The abscess may gravitate into the thigh via the femoral vascular sheath or present in the buttock via the greater or the lesser sciatic notch. Ischiorectal presentation may occur. Retroperitoneal extension into pelvic organs also has been reported. The 3rd common route is through a single sheet of lumbodorsal fascia into Petit's triangle, lateral to the point where the lumbodorsal fascia splits into 3 layers. Figures 2 and 3 clarify the anatomic pathways described.

*Annie C. Kane Fellow, New York Orthopaedic Hospital, Columbia-Presbyterian Medical Center, New York, N. Y.

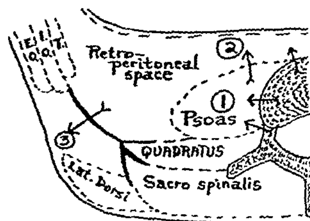


FIG. 1. Pathways tracked by dorsolumbar tuberculous abscesses: (1) within psoas sheath; (2) retroperitoneal; (3) lumbar (Petit's) triangle.

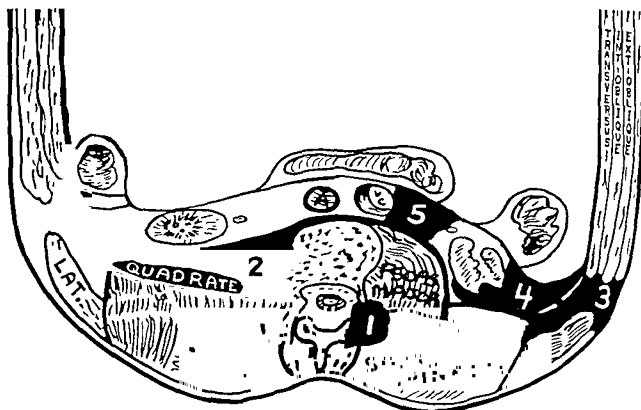


FIG. 2. Abscess sites: (1) vertebral focus; (2) within psoas sheath; (3) lumbar triangle; (4) perirenal; (5) retroperitoneal. (After Callander, C. L.: Surgical Anatomy, ed. 3, Philadelphia, Saunders)

REPRESENTATIVE CASE REPORTS

1. Pathway: Within Psoas Sheath. A 48-year-old Negro. Entered the New York Orthopaedic Hospital in 1952 with fatigue and moderate weight loss of 6 months' duration and left hip pain of 4 months' duration. Roentgenograms demonstrated a destructive lesion of D12-L1 with obliteration of the left psoas shadow. A mass was present in the left iliac fossa, which was incised and drained after the patient had been placed on antituberculous drugs. A smear from abscess material revealed acid-fast bacilli. A long rubber catheter was threaded into the iliac abscess and passed cephalad. Roentgenograms were taken demonstrating the catheter passing up the psoas sheath to the side of the bodies of D12-L1 (Fig. 4). Late in 1953, a spinal fusion, D11-L2 was performed Today the patient is alive and well.



FIG 3. Abscess within psoas sheath on left. Retroperitoneal abscess, presenting in buttock via sciatic notch on right (after Calot).

2. Pathway: Retroperitoneal into Buttock via Sciatic Notch. A 43-year-old Negro. Entered the New York Orthopaedic Hospital in 1953 with urinary frequency and nocturia of 5 years' duration and a left thigh mass of 4 months' duration. He sought medical advice because the size of the left thigh and buttock mass interfered with his passing through subway turnstiles. Roentgenograms revealed a destruc-



FIG. 4. 48-year-old Negro. Tuberculosis D12-L1. A catheter was passed cephalad through iliac fossa abscess via psoas sheath to vertebral focus.

tive lesion of L3-L4. Aspiration of the abscess was performed, and acid-fast bacilli were demonstrated on smear. A total of 3,050 cc. of thick gray-green flocculent material was aspirated from the abscess over a 2-month period. The patient, who was receiving antituberculous drugs, never developed a sinus tract. Radiopaque dye was injected into the buttock abscess but apparently failed to pass through the sciatic notch into the retroperitoneal space because of fibrous stenosis of the communicating pathway (Fig 5). A spinal fusion L2-L5 was done in 1953. Today the patient is alive and well.

3. Pathway: Retroperitoneal into Both Thighs via Vascular Sheaths. A 27-year-old Negress. Entered the New York Orthopaedic Hospital in 1951 with low back pain of 6 months' duration. Roentgenograms revealed a destructive lesion of the right side of the sacrum and the body of L5. Biopsy of the right side of sacrum revealed histologic evidence of tuberculosis. A left thigh abscess was present which, when aspirated, yielded 4,800 cc. of thick gray-green flocculent material. Radiopaque dye injected into the abscess cavity passed through the retroperitoneal space into the opposite thigh (Fig. 6). A left hemifusion L4-S1 was performed in 1951 and was supplemented by right sacro-iliac and



FIG. 2. Abscess sites: (1) vertebral focus; (2) within psoas sheath; (3) lumbar triangle; (4) perirenal; (5) retroperitoneal. (After Callander, C. L.: Surgical Anatomy, ed. 3, Philadelphia, Saunders)

REPRESENTATIVE CASE REPORTS

1. Pathway: Within Psoas Sheath. A 48-year-old Negro. Entered the New York Orthopaedic Hospital in 1952 with fatigue and moderate weight loss of 6 months' duration and left hip pain of 4 months' duration. Roentgenograms demonstrated a destructive lesion of D12-L1 with obliteration of the left psoas shadow. A mass was present in the left iliac fossa, which was incised and drained after the patient had been placed on antituberculous drugs. A smear from abscess material revealed acid-fast bacilli. A long rubber catheter was threaded into the iliac abscess and passed cephalad. Roentgenograms were taken demonstrating the catheter passing up the psoas sheath to the side of the bodies of D12-L1 (Fig. 4). Late in 1953, a spinal fusion, D11-L2 was performed. Today the patient is alive and well.



FIG. 3. Abscess within psoas sheath on left. Retroperitoneal abscess, presenting in buttock via sciatic notch on right (after Calot).

2. Pathway: Retroperitoneal into Buttock via Sciatic Notch. A 43-year-old Negro. Entered the New York Orthopaedic Hospital in 1953 with urinary frequency and nocturia of 5 years' duration and a left thigh mass of 4 months' duration. He sought medical advice because the size of the left thigh and buttock mass interfered with his passing through subway turnstiles. Roentgenograms revealed a destruc-



FIG. 4. 48-year-old Negro. Tuberculosis D12-L1. A catheter was passed cephalad through iliac fossa abscess via psoas sheath to vertebral focus.

tive lesion of L3-L4. Aspiration of the abscess was performed, and acid-fast bacilli were demonstrated on smear. A total of 3,050 cc. of thick gray-green flocculent material was aspirated from the abscess over a 2-month period. The patient, who was receiving antituberculous drugs, never developed a sinus tract. Radiopaque dye was injected into the buttock abscess but apparently failed to pass through the sciatic notch into the retroperitoneal space because of fibrous stenosis of the communicating pathway (Fig. 5). A spinal fusion L2-L5 was done in 1953. Today the patient is alive *and well.*

3. Pathway: *Retroperitoneal into* Both Thighs via Vascular Sheaths. A 27-year-old Negress. Entered the New York Orthopaedic Hospital in 1951 with low back pain of 6 months' duration. Roentgenograms revealed a destructive lesion of the right side of the sacrum and the body of L5. Biopsy of the right side of sacrum revealed histologic evidence of tuberculosis. A left thigh abscess was present which, when aspirated, yielded 4,800 cc. of thick gray-green flocculent material. Radiopaque dye injected into the abscess cavity passed through the retroperitoneal space into the opposite thigh (Fig. 6). A left hemifusion L4-S1 was performed in 1951 and was supplemented by right sacro-iliac and

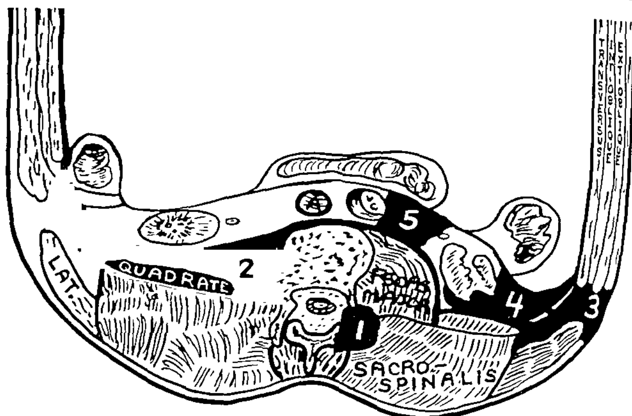


FIG. 2. Abscess sites: (1) vertebral focus; (2) within psoas sheath; (3) lumbar triangle; (4) perirenal; (5) retroperitoneal. (After Callander, C. L.: Surgical Anatomy, ed. 3, Philadelphia, Saunders)

REPRESENTATIVE CASE REPORTS

1. Pathway: Within Psoas Sheath. A 48-year-old Negro. Entered the New York Orthopaedic Hospital in 1952 with fatigue and moderate weight loss of 6 months' duration and left hip pain of 4 months' duration. Roentgenograms demonstrated a destructive lesion of D12-L1 with obliteration of the left psoas shadow. A mass was present in the left iliac fossa, which was incised and drained after the patient had been placed on antituberculous drugs. A smear from abscess material revealed acid-fast bacilli A long rubber catheter was threaded into the iliac abscess and passed cephalad. Roentgenograms were taken demonstrating the catheter passing up the psoas sheath to the side of the bodies of D12-L1 (Fig. 4). Late in 1953, a spinal fusion, D11-L2 was performed. Today the patient is alive and well.

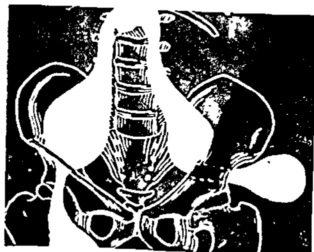


FIG. 3. Abscess within psoas sheath on left. Retroperitoneal abscess, presenting in buttock via sciatic notch on right (after Calot).

2. Pathway: Retroperitoneal into Buttock via Sciatic Notch. A 43-year-old Negro. Entered the New York Orthopaedic Hospital in 1953 with urinary frequency and nocturia of 5 years' duration and a left thigh mass of 4 months' duration. He sought medical advice because the size of the left thigh and buttock mass interfered with his passing through subway turnstiles. Roentgenograms revealed a destruc-



FIG. 4. 48-year-old Negro. Tuberculosis D12-L1. A catheter was passed cephalad through iliac fossa abscess via psoas sheath to vertebral focus.

tive lesion of L3-L4. Aspiration of the abscess was performed, and acid-fast bacilli were demonstrated on smear. A total of 3,050 cc. of thick gray-green flocculent material was aspirated from the abscess over a 2-month period. The patient, who was receiving antituberculous drugs, never developed a sinus tract. Radiopaque dye was injected into the buttock abscess but apparently failed to pass through the sciatic notch into the retroperitoneal space because of fibrous stenosis of the communicating pathway (Fig. 5). A spinal fusion L2-L5 was done in 1953. Today the patient is alive and well.

3. **Pathway: Retroperitoneal into Both Thighs via Vascular Sheaths.** A 27-year-old Negress. Entered the New York Orthopaedic Hospital in 1951 with low back pain of 6 months' duration. Roentgenograms revealed a destructive lesion of the right side of the sacrum and the body of L5. Biopsy of the right side of sacrum revealed histologic evidence of tuberculosis. A left thigh abscess was present which, when aspirated, yielded 4,800 cc. of thick gray-green flocculent material. Radiopaque dye injected into the abscess cavity passed through the retroperitoneal space into the opposite thigh (Fig. 6). A left hemifusion L4-S1 was performed in 1951 and was supplemented by right sacro-iliac and





FIG. 5. 43-year-old Negro. (*Top, left, and bottom*) Tuberculosis L3-L4. Large buttock abscess via retroperitoneal route. (*Top, right*) Dye in buttock abscess.



right L5-S1 fusions in 1953. The patient is now well without complaints.

4. Pathway: Lumbar (Petit's) Triangle. A 6-year-old white female. Entered the New York Orthopaedic Hospital in 1943 with progressive dorsal kyphosis of 2 years' duration and a large nontender mass in the right lumbar area. Aspiration of this abscess was followed by a positive culture for acid-fast bacilli. Roentgenograms revealed a destructive vertebral focus at D12-L1 (Fig. 7). A spinal fusion D9-L4 was performed in 1943. Ten years later, the patient was symptom-free without evidence of active disease.

SUMMARY

The common anatomic pathways tracked by tuberculous abscesses from the lower dorsal and lumbar spine have been described. Representative case reports from the New York Orthopaedic Hospital, with appropriate roentgenograms and clinical photographs, have been included, illustrating these pathways.

REFERENCES

1. Hallock, H., and Jones, J. B.: Tuberculosis of the spine, J. Bone & Joint Surg. 36A: 219-241, 1954.
2. Mercer, W.: Orthopaedic Surgery, ed. 4, pp. 262-311, Baltimore, Williams & Wilkins, 1950.
3. Steindler, A.: Postgraduate Lectures on Orthopedic Diagnosis and Indications, vol. 3, pp. 27-59, Springfield, Ill., Thomas, 1952.

Trajectos Sequite per Tuberculotic Abscessos Dorso-Lumbar

Summario in Interlingua

Depost le advento de potente drogas antituberculotic, le diagnose e le subsequente tractamento de paravertebral abscessos tuberculotic ha experientiate grande alterationes. Indolente trajectos sinusal ha practicamente disparite. Al Hospital Orthopedic New York, un numero de tuberculotic abscessos originari del spina dorso-lumbar es-



Fig. 6. 27-year-old Negress. (*Top, left and right*) Tuberculosis of L5 and right side of sacrum. Large thigh abscess via vascular sheaths from retroperitoneal space. (*Bottom, left*) Dye in large thigh abscess. (*Bottom, right*) Dye injected into large thigh abscess has extended, via retroperitoneal space, into opposite thigh.



FIG. 7. 6-year-old white female. (*Left*) Tuberculosis D12-L1: large lumbar abscess. (*Right*) Tuberculosis D12-L1. (Hallock & Jones: J. Bone & Joint Surg. 36A:221 & 222)

seva recentemente studiate per medio del injection directe de radio-opac substantias colorante. Le resultato esseva un vivissime demonstration del trajectos anatomic sequite per iste abscessos.

Le frequentia de "frigide" abscessos associate con tuberculose vertebral varia ab 20 pro cento[1] a 63 pro cento.[2] Apparentemente iste sequela es plus frequente in morbo infero-dorsal e lumbar que in affectiones del altere segmentos spinal. In un serie recentemente reportate ab iste instituto, Hallock e Jones[3] trovava que 91 pro cento del casos de tuberculose spinal ha lor origine in le segmentos infero-dorsal, dorso-lumbar, e lumbar.

Trajectos Anatomic. Figura 1 delinea le tres commun trajectos sequite per abscessos partiente ab le spina infero-dorsal o lumbar. Forsan le plus frequente via progrede intra le vaina del musculo psoatic, con presentation al fossa iliac o gravitation a basso usque a infra le ligamento inguinal, usualmente al aspecto adductori del coxa. Le abscesso pote penetrar (1) le anterior ligamento longitudinal del spina o (2) le vaina psoatic ventral pro obtener accesso al spatio retroperitoneal. Postea plure vias es possibile. Le abscesso pote gravitar a in le femore via le vaina vascular femoral o presentar se al nates via le major o minor incisura sciatic. Presentation ischio-rectal es possibile. Etiam extensiones retroperitoneal a in le organos pelvic ha essite reportate. Le tertie via commun progrede a transverso un sol fascia lumbo-dorsal verso le triangulo de Petit, al latere del puncto ubi le fascias lumbo-dorsal se separa in tres stratos. Figura 2 e figura 3 illustra le trajectos anatomic describite.

Es describite le commun trajectos anatomic sequite per abscessos tuberculotic partiente ab le spina infero-dorsal e lumbar. Es includite, como illustrationes de ille trajectos, reportos de casos representative ab le archivos del Hospital Orthopedic New York, accompaniate de appropriate roentgenogrammas e photographias clinic.

22

The Jan Zahradnicek Surgical Approach to the Problem of Congenital Hip Dislocation

J. E. M. THOMSON, M.D.*

The experiences of Professor Jan Zahradnicek, while chief of the First Clinic for Orthopaedic Surgery of Charles University in Prague, with the operative treatment of congenital dislocation of the hip has been so extensive and well known in Europe that it deserves wider dissemination in English literature.

Having had the opportunity of working for some time in the Prague clinic and seeing the excellent precision technic and results, as well as the enthusiastic response to the professor's work of his devoted students, assistants, followers and patients, one cannot help being impressed and having the highest respect for his accomplishments. It is the purpose of the author in this brief chapter to stimulate others to consider his ideas, which have had such far-reaching influence in Central-European countries.

The incidence of congenital dislocation of the hip in Central Europe is great, and the increase was significant during the period of World War II and shortly afterward. The orthopaedic surgeons of these countries long have recognized the shortcomings of the conventional Lorenz and modified forcible reductions of congenital dislocation of the hip followed by long periods of restriction in plaster of Paris casts. True, the early results of many were quite good But at puberty, quite a number of these patients developed a limp and instability. At 20 years of age, there were more with a limp, and at 40 nearly all of those regarded early as good results had distinct disability in the hip. Their feeling is that only about 10 per cent of those treated by the manual reduction and plaster method showed satisfactory results.

The gradual reduction of the dislocation, by the use of the pillow apparatus of Frejka and the biomechanical apparatus of Hanausek used in more stubborn cases, permitted accomplishment of the reduction without trauma and, as a result, with much better functional outlook in later life.

The discussion of these methods is beyond the scope of this chapter. However, there were many cases that did not respond to these gradual conservative reduction methods. This fact led Professor Zahradnicek to study the factors that influenced these failures. His conclusions are that to have a permanent functional success in the treatment of congenital dislocation of the hip, the normal anatomic relationships must be re-established between the acetabulum and the head and the neck of the femur and the shaft. Further, the head must be centered adequately in a well-formed acetabulum; the angle of the neck should be about 125° (not much greater or not much less); and anteversion must be corrected accurately. It is the purpose of the surgical operations devised by Professor Zahradnicek to accomplish a restoration of normal hip relationship.

* Lincoln, Neb.

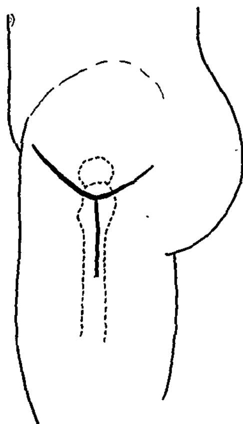


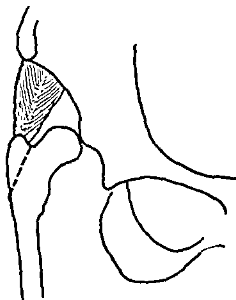
FIG. 1. The T-incision for approach to the hip carried *from below the anterior superior spine downward* across the greater trochanter, back and upward toward the tubercle of the ischium. Then the vertical limb of the T begins over the center of the trochanter and extends downward. The length is determined by the extent of the procedure.

The ideal time for operations is from 3 to 6 years of age. The earlier age is advisable when conservative measures have shown acknowledged failure. The older age is reserved for those in whom the pathology is not recognized or for those who were neglected. Beyond the age of 6 years these procedures are not advised.

TECHNIC

The Zahradnicek approach to the hip comprises *a T-shaped skin incision (Fig. 1)* performed with the patient lying on the healthy side and the dislocated trochanter and femur pointing directly upward toward *the surgeon.*

The horizontal limb of the T is a curved incision, extending from below the anterior superior spine of the ilium downward over the greater trochanter, backward and upward toward the tuberosity of the ischium. The vertical limb of the T begins over the center of the trochanter and extends inferiorly. The length of this latter incision is determined by the type and the extent of operation planned for the particular case. The horizontal incision follows somewhat



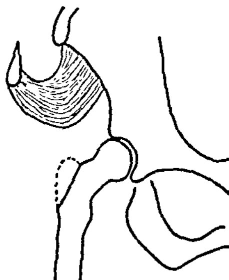


FIG. 2 (*Left*) A long oblique pretrochanteric osteotomy is performed.
FIG. 3 (*Right*) The soft tissue and the muscle are reflected upward and back *with the trochanter.*

the anterior border of the tensor fascia lata downward across the trochanter and backward across the mid-gluteus maximus; the fascia lata is cut transversely at the level of the skin incision. The skin is reflected over the trochanter. A long oblique pretrochanteric osteotomy is performed, carrying as much of the upper insertion of the gluteus maximus and all of the gluteus medius and minimus (Fig. 2). This osteotomy should be as long and as oblique as possible, and the soft tissues and the muscles are reflected upward and backward with the osteotomized greater trochanter (Fig. 3). This procedure exposes completely the hip joint and the upper end of the femur and gives a splendid vision for the correction of coxa valga, coxa vara and anteversion; it also provides an opportunity to ensure a well-formed acetabulum for the proper centering of the head of the femur. Great emphasis is made on centering the head in a well-formed acetabulum.

Having a view of the pathology to be dealt with, the following methods are used for correction of the various deformities:

Should the common pathology of coxa valga exist, a triangle wedge through the trochanter is made with the base near the lesser trochanter (Fig. 4, *left*). The size of the wedge depends upon the degree of correction desired. After the wedge is removed and the deformity has been corrected, a flanged nail similar to, but smaller than, a Smith-Petersen nail is used to immobilize the fragments after correction (Fig. 4, *right*).

The wedge that is removed may be used as a shelf over the acetabulum if this is deemed necessary to give the hip better stability. The greater trochanter is brought down and held by screws or small nails, and the patient is placed in a single spica cast.

If the deformity with which we are dealing is one of coxa vara, the triangular wedge is taken in the opposite direction with the apex of the lesser trochanter and the base through the great trochanter, the angle and the size estimated to restore the neck to the 125° angle. After correction, again a flanged

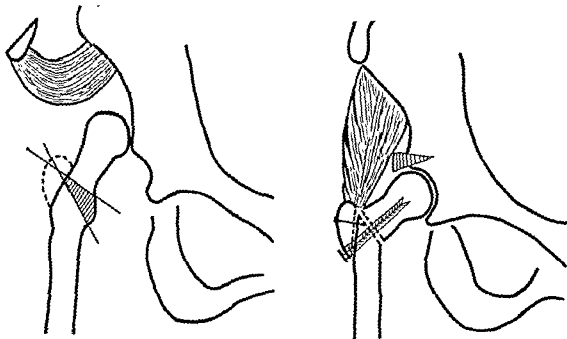


FIG. 4. (*Left*) Congenital dislocation with coxa valga. The wedge-shaped shaded area is removed with the base downward. (*Right*) The wedge-shaped area that is removed may be used as a shelf above the acetabulum if needed. The head is reduced into the acetabulum, and the corrected neck is stabilized with a flanged nail. Also, a screw or a nail is used to secure the trochanter and the glutei in place.

nail is inserted to maintain the fragments in position. Again the wedge may be used as before and the trochanter fixed in place, as described previously (see Fig. 5).

In most of these cases of coxa valga or coxa vara, there is a degree of anteversion (Fig. 6) that must be considered in acquiring the triangular wedge. The angle of the wedge must be established so that when removed, correction of this deformity can be accomplished. However, in some of these cases it is desirable to correct the anteversion before dealing with any other deformity. In the correction of anteversion, the wedge must be made with the base of the triangle backward and downward. Figures 7 and 8 show the procedure diagrammatically.

We have long recognized the inadvisability of endeavoring to reduce the hip into the original acetabulum when the head has long been riding high on the ilium. Professor Zahradnicek solves the problem of bringing the hip down to its original acetabulum and reams out an adequate seat or shelf above to ensure stability when the head is reduced. If the head is forced into this corrected relationship with undue pressure from muscles, Professor Zahradnicek and his workers have shown that not only will disability be ensured and constant but also aseptic necrosis of the head usually occurs. For that reason he believes that the femur should be shortened sufficiently so that the reconstructed head and neck in the acetabulum are not under stress or pressure after correction. For that reason he takes an oblique section out of the femur near the greater trochanter, which will ensure the 125° angle of the shaft with the neck and again stabilize the fragments with a flanged nail with, at times, circumscribing stainless steel wires (Fig. 9).

In all these corrective procedures the excess capsule and debris of the acetabulum are removed.

AFTER-CARE

A spica cast is applied from the ribs to the toes in mild abduction, and the child remains in this cast for 18 days The ex-

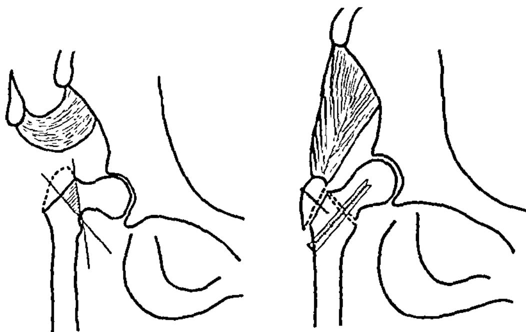


Fig 5. (*Left*) Should the hip have a coxa vara deformity, the wedge is taken with the base above (*Right*) The wedge removed and deformity of the neck corrected and stabilized by flanged nail. The trochanter attached by a screw or a nail.

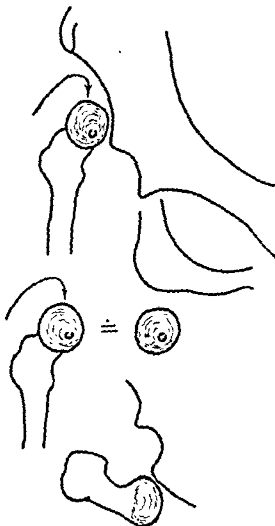


FIG. 6. (*Top*) A congenital dislocation of the hip with anteversion of the neck of the femur. (*Center*) Schematic appearance of the head (*Bottom*) Looking from above downward in cross section the head is ventral and above in relationship to the acetabulum.

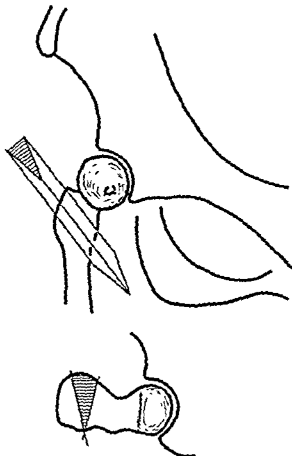


FIG. 7. (*Top*) In the correction of anteversion the wedge has its base to the back. (*Bottom*) Cross section of neck and head showing the segment of the wedge removed.

perience at Prague has led to the belief that by this time sufficient adhesive bony regeneration has taken place in the oblique osteotomies that have been fixed with the flanged nail to allow the patient to be in bed with mild traction and balance support. Exercises and movement now are encouraged, and physical therapy can be started. This bed treatment is continued for about 2 weeks Then the patient is allowed to be out of bed and to walk on crutches. Full weight-bearing without crutches should not be started for from 3 to 6 months, depending upon the roentgen evidence of good healing and result.

It is impossible for me to obtain accurate figures of results, but, up to a few years ago, Professor Zahradnicek and his associates had performed from 100 to 150 such operations each year, and the results that I was privileged to examine, as the patients appeared for routine checkups from 1 to 8 years after operation, seemed far more satisfactory than many that I have seen in other clinics.

Our modest experience with these technics is not sufficient to justify personal conclusions. To many, these procedures may seem far afield from the orthodox concept of this problem. However, there is much to incite the imagination and provoke controversy

nail is inserted to maintain the fragments in position. Again the wedge may be used as before and the trochanter fixed in place, as described previously (see Fig. 5).

In most of these cases of coxa valga or coxa vara, there is a degree of anteversion (Fig. 6) that must be considered in acquiring the triangular wedge. The angle of the wedge must be established so that when removed, correction of this deformity can be accomplished. However, in some of these cases it is desirable to correct the anteversion before dealing with any other deformity. In the correction of anteversion, the wedge must be made with the base of the triangle backward and downward. Figures 7 and 8 show the procedure diagrammatically.

We have long recognized the inadvisability of endeavoring to reduce the hip into the original acetabulum when the head has long been riding high on the ilium. Professor Zahradnicek solves the problem of bringing the hip down to its original acetabulum and reams out an adequate seat or shelf above to ensure stability when the head is reduced. If the head is forced into this corrected relationship with undue pressure from muscles, Professor Zahradnicek and his workers have shown that not only will disability be ensured and constant but also aseptic necrosis of the head usually occurs. For that reason he believes that the femur should be shortened sufficiently so that the reconstructed head and neck in the acetabulum are not under stress or pressure after correction. For that reason he takes an oblique section out of the femur near the greater trochanter, which will ensure the 125° angle of the shaft with the neck and again stabilize the fragments with a flanged nail with, at times, circumscribing stainless steel wires (Fig. 9).

In all these corrective procedures the excess capsule and debris of the acetabulum are removed.

AFTER-CARE

A spica cast is applied from the ribs to the toes in mild abduction, and the child remains in this cast for 18 days. The ex-

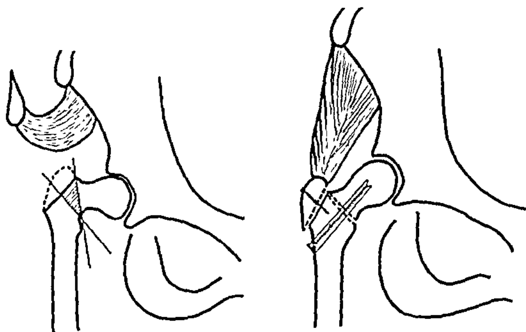


Fig. 5. (*Left*) Should the hip have a coxa vara deformity, the wedge is taken with the base above. (*Right*) The wedge removed and deformity of the neck corrected and stabilized by flanged nail. The trochanter attached by a screw or a nail.

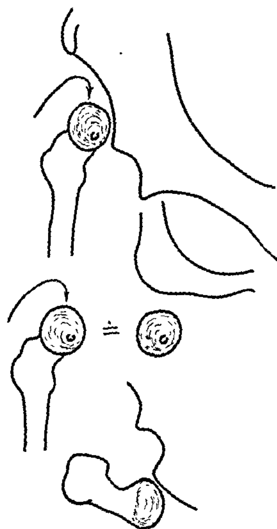


Fig. 6. (*Top*) A congenital dislocation of the hip with anteversion of the neck of the femur. (*Center*) Schematic appearance of the head. (*Bottom*) Looking from above downward in cross section the head is ventral and above in relationship to the acetabulum.

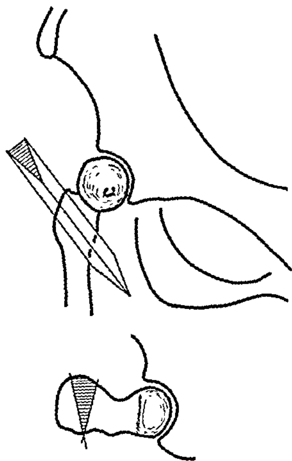


Fig. 7. (*Top*) In the correction of anteversion the wedge has its base to the back. (*Bottom*) Cross section of neck and head showing the segment of the wedge removed.

perience at Prague has led to the belief that by this time sufficient adhesive bony regeneration has taken place in the oblique osteotomies that have been fixed with the flanged nail to allow the patient to be in bed with mild traction and balance support. Exercises and movement now are encouraged, and physical therapy can be started. This bed treatment is continued for about 2 weeks. Then the patient is allowed to be out of bed and to walk on crutches. Full weight-bearing without crutches should not be started for from 3 to 6 months, depending upon the roentgen evidence of good healing and result.

It is impossible for me to obtain accurate figures of results, but, up to a few years ago, Professor Zahradnicek and his associates had performed from 100 to 150 such operations each year, and the results that I was privileged to examine, as the patients appeared for routine checkups from 1 to 8 years after operation, seemed far more satisfactory than many that I have seen in other clinics.

Our modest experience with these technics is not sufficient to justify personal conclusions. To many, these procedures may seem far afield from the orthodox concept of this problem. However, there is much to incite the imagination and provoke controversy

concerning the possibilities for the future of patients so treated.

Dr. Vaclav Tosovsky and Dr. Jaroslav Slavik, of the Zaharadnicek Clinic, who have helped in the preparation of this material, emphasize the following salient points in explaining the reasons for their results:

1. The age of the patient is limited to those between 3 and 6 years.
2. Surgical exploration and correction *only* can restore function.
3. This approach to the joint is less damaging to vital circulation than any other.
4. Osteotomy and corrective wedges through the trochanteric region with rich blood supply tend to rapid bone healing.
5. Centering the head of the femur in the natural habitat of the normal acetabular position, even at the expense of shortening the extremity, is a factor in ensuring a measure of good function.
6. Re-establishment of circulating equilibrium in the head is more likely to be accomplished readily under the above conditions of age, approach and corrective measures.

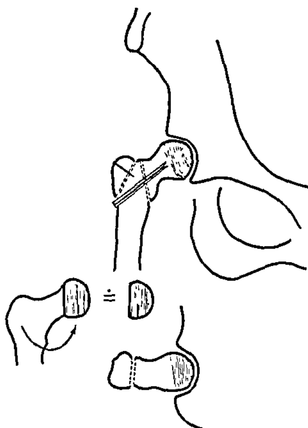


FIG. 8. (*Top*) After correction of anteversion. (*Center*) Schematic relationship of the head and the neck after correction. (*Bottom*) Cross section after correction of the anteversion.



FIG. 9. (*Left*) Professor Zahradnicek believes that in older children it is preferable to shorten the leg by means of removal of an oblique section of the appropriate length just below the trochanters and reduce the heads into the site of the original acetabulum. The acetabulum must be adequate or reamed out to give good centering of the head (*Right*) After reducing the head and shortening the femur the flanged nail is used to stabilize the osteotomy.

Le Problema del Congenite Dislocation Coxal e su Resolution Chirurgic Secundo le Methodo de Jan Zahradnicek

Summario in Interlingua

Le labores de professor Zahradnicek in le area del tractamento chirurgic de congenite dislocation coxal es si extense e si ben cognoscite in Europa que illos merita un plus adequate representation in le litteratura de lingua anglese.

Zahradnicek e altere chirurgos de Europa Central recognosceva le defectos del technica conventional de reduction forciate, i.e. le operation de Lorenz e le modificationes de illo, in tanto que le resultatos favorabile amontava solmente a 10 pro cento. Per consequente, professor Zahradnicek se sentiva inducite a studiar le factores que causa tante insuccessos. Su conclusion esseva que un permanente successo functional require le restablimento del normal relation anatomic inter acetabulo, capite, colo, e diaphyse del femore. Le sequente es le considerationes principal que explica le bon successo del methodo de Zahradnicek.

1. Le tempore ideal pro le operation es inter le tertie e le sexte anno del etate del patiente.

2. Exploration e correction chirurgic es indispensabile pro restaurar le functiones.

3. Le operation de Zahradnicek es minus damnose al circulation vital que altere operationes.

4. Rapide curation del osso es assecurate per render le osteotomia le plus longe e le plus oblique possibile, durante que un vision perfecte es obtenite per le reflexion del musculo in alto e in retro, e per placiar le cuneos corrective a transverso le region trochanteric que es ric in apporto de sanguine. In certe casos, il es ver, on debe corriger le anteversion in preparation al correction del deformitate.

5. Bon effectos functional es obtenite per le placiamento central del capite femoral in un normal position acetabular, mesmo si isto resulta in un accurtamento del extremitate.

6. Le restablimento del equilibrio circulatori in le capite femoral es plus probabile sub le supra-listate conditiones de etate del patiente e de mesurase corrective usate per le chirurgo.

concerning the possibilities for the future of patients so treated.

Dr. Vaclav Tosovsky and Dr. Jaroslav Slavik, of the Zaharadnicek Clinic, who have helped in the preparation of this material, emphasize the following salient points in explaining the reasons for their results:

1. The age of the patient is limited to those between 3 and 6 years.
2. Surgical exploration and correction *only* can restore function.
3. This approach to the joint is less damaging to vital circulation than any other.
4. Osteotomy and corrective wedges through the trochanteric region with rich blood supply tend to rapid bone healing.
5. Centering the head of the femur in the natural habitat of the normal acetabular position, even at the expense of shortening the extremity, is a factor in ensuring a measure of good function.
6. Re-establishment of circulating equilibrium in the head is more likely to be accomplished readily under the above conditions of age, approach and corrective measures.

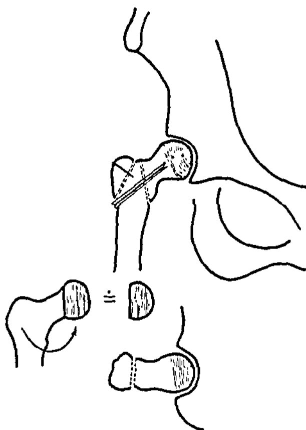


Fig. 8. (*Top*) After correction of anteversion. (*Center*) Schematic relationship of the head and the neck after correction. (*Bottom*) Cross section after correction of the anteversion.

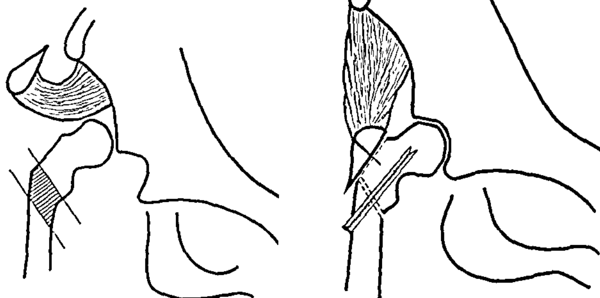


Fig. 9. (*Left*) Professor Zahradnicek believes that in older children it is preferable to shorten the leg by means of removal of an oblique section of the appropriate length just below the trochanters and reduce the heads into the site of the original acetabulum. The acetabulum must be adequate or reamed out to give good centering of the head. (*Right*) After reducing the head and shortening the femur the flanged nail is used to stabilize the osteotomy

Le Problema del Congenite Dislocation Coxal e su Resolution Chirurgic Secundo le Methodo de Jan Zahradnicek

Summario in Interlingua

Le labores de professor Zahradnicek in le area del tractamento chirurgic de congenite dislocation coxal es si extense e si ben cognoscite in Europa que illos merita un plus adequate representation in le litteratura de lingua anglese.

Zahradnicek e altere chirurgos de Europa Central recognosceva le defectos del technica conventional de reduction forciate, i.e. le operation de Lorenz e le modificationes de illo, in tanto que le resultatos favorabile amontava solmente a 10 pro cento. Per consequente, professor Zahradnicek se sentiva inducite a studiar le factores que causa tante insuccessos. Su conclusion esseva que un permanente successo functional require le restablimento del normal relation anatomic inter acetabulo, capite, colo, e diaphyse del femore. Le sequente es le considerationes principal que explica le bon successo del methodo de Zahradnicek.

1. Le tempore ideal pro le operation es inter le tertie e le sexte anno del etate del patiente.

2. Exploration e correction chirurgic es indispensabile pro restaurar le functiones.

3. Le operation de Zahradnicek es minus damnose al circulation vital que altere operationes.

4. Rapide curation del osso es assecurate per render le osteotomía le plus longe e le plus oblique possibile, durante que un vision perfecte es obtenite per le reflexion del musculo in alto e in retro, e per placiar le cuneos corrective a transverso le region trochanteric que es ric in apporto de sanguine. In certe casos, il es ver, on debe corriger le anteversion in preparation al correction del deformitate.

5. Bon effectos functional es obtenite per le placiamento central del capite femoral in un normal position acetabular, mesmo si isto resulta in un accurtamento del extremitate.

6. Le restablimento del equilibrio circulatori in le capite femoral es plus probabile sub le supra-listate conditiones de etate del patiente e de mesurase corrective usate per le chirurgo.

23

Carpometacarpal Dislocation

A Case Report

DONALD T. JONES, M.D.,* AND CHESTER W. ESKEY, M.D.*

Carpometacarpal dislocation of any one or any combination of the medial 4 metacarpals is a relatively rare injury. In view of this we feel justified in reporting this single case.

Dislocation of all 4 medial metacarpals is the most usual combination. Next in frequency is isolated dislocation of the 5th metacarpal, followed by dislocation of metacarpals 4 and 5 in combination. There are 10 reported cases of dislocation of the 2nd and the 3rd metacarpals; this case is the 11th. Of the remaining combinations or isolated dislocations there are fewer than 10 cases of any one type reported. Waugh and Yancey in 1948 have tabulated these and the dates of the initial reports in an excellent extensively documented report. More recently, Whitson has reviewed these and added another case of posterior dislocation of all 4 medial metacarpals. The usual displacement is dorsal. In reviewing cases of dislocation of the 2nd and the 3rd metacarpals simultaneously, only one previous volar dislocation was discovered

CASE REPORT

The patient, a 21-year-old student, was driving a tractor towing a steel girder. The girder became caught, and the tractor reared up. The patient dived off, attempting to get clear. However, the rear wheel rolled across his chest, leaving a clear path of abrasions from the anterior aspect of his right shoulder across his sternum ending just below the nipple line on the left. He does not remember the tractor wheels rolling over his hand.

The patient was seen within one half hour following his injury. Examination and roentgenograms on admission and subsequently, with the exception of his right hand, failed to show any injuries other than extensive contusions and abrasions, although fractures of the sternum, the left radius and posterior dislocation of his left shoulder were suspected at the time of his admission.

The right hand initially showed a hematoma over the dorsal aspect of the 2nd and the 3rd metacarpals and a palpable prominence in the palm just medial to the thenar eminence. There were no abrasions of the skin of the hand to suggest direct trauma. The swelling over the dorsum enlarged perceptibly, and the palm became very tense during the time required to take the roentgenograms. The films revealed a fracture of the distal phalanx of the 2nd finger and volar dislocation of the 2nd and the 3rd metacarpals with subluxation of the 4th metacarpal in a radial direction. Ten cc. of 1 per cent procaine and 600 turbidity units of hyaluronidase were injected from the dorsal aspect into the palpable defect just distal to the distal carpal row. Compression was applied with an Ace bandage and man-

* Philadelphia, Pa.

ually for a period of 10 minutes. This gave satisfactory anesthesia. Strong traction on the 2nd and the 3rd fingers, coupled with pressure over the metacarpal bases, reduced the dislocation with an audible and palpable crepitation. Postreduction roentgenograms showed satisfactory reduction. The hand was immobilized in flexion, the splints extending from the fingertips to the elbow. Two weeks following injury the cast was shortened to the distal palmar crease, and finger motion was encouraged. The cast was removed five weeks following reduction. Follow-up roentgenograms have shown maintenance of the reduction, and finger motion has been improving steadily.

The rarity of these lesions, especially in this industrialized age in which the hand is subjected to such frequent and violent trauma, testifies to the stability of the carpometacarpal articulations of the medial 4 metacarpals. The joints between the medial 4 metacarpals and the distal carpal row are modified saddle joints. These joints derive appreciable stability from both their bony configuration and their supporting ligaments. The 2nd and the 3rd metacarpals are practically immobile on the distal carpal row with the 4th metacarpal having slightly more motion and the 5th metacarpal the greatest amount. This motion is largely flexion and extension. The distal carpal row presents multiple irregular articulating surfaces for the 4 medial metacarpals. The 2nd metacarpal is morticed securely between the greater multangular and the capitate and an anteroposteriorly directed ridge on the lesser multangular fits securely into the proximal end of the 2nd metacarpal. This bony stability of the 2nd metacarpal largely accounts for the stability of the remaining carpometacarpal articulations. It particularly controls radial or ulnar shift of the medial 4 metacarpals. Metacarpals 4 and 5 are permitted some ulnar deviation by the sloping distal surface of the hamate. The articular

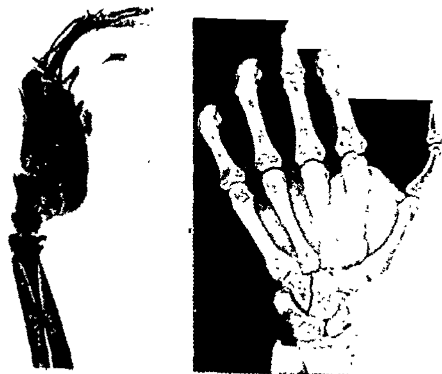


Fig. 1. Roentgenograms on admission to hospital show volar dislocation of the 2nd and the 3rd metacarpals on the carpus and subluxation of the 4th metacarpal in a radial direction; also comminution about the proximal ends of the metacarpals and a fracture of the distal phalanx 2nd finger.

capsule of the carpometacarpal joints unites with the intercarpal capsules and is taut with the exception of the 5th metacarpal. The dorsal carpometacarpal ligaments and the volar carpometacarpal ligaments, along with the interosseous carpometacarpal ligament, further enhance the stability. The articular cavity is a continuation of the synovial cavity of the intercarpal joints. In some instances the articulation between the hamate and the 4th and the 5th metacarpals does not communicate with the common joint cavity

Both indirect and direct trauma have been recorded as causing carpometacarpal dislocation.

Experimental production of these injuries on cadavers by Shorbe was possible only after division of the dorsal and volar ligaments. Following division of the ligaments the bases could be dislocated under direct pressure with the hand in acute flexion. Biacheri has noted the frequent occurrence of laceration of the dorsal ligaments following forceful flexion. Recurrence of dorsal dislocation when splinted in flexion has been reported by several authors. Bannes reports a dorsal dislocation of the 2nd and the 3rd metacarpal following trauma over the dorsal aspect of the distal end, apparently indirectly levering out the bases. A similar mechanism is reported by Humbert and Berdach for dorsal dislocation of the 2nd metacarpal alone. However, direct trauma accounts for probably the greatest number of cases. Whitson gives as an example of the mechanism for dorsal dislocation the sudden arrest of momentum of someone grasping a steering wheel or handlebars. Orrillard cites the case of a metal worker who was holding his hand over a depression and sustained direct trauma over the dorsum with subse-

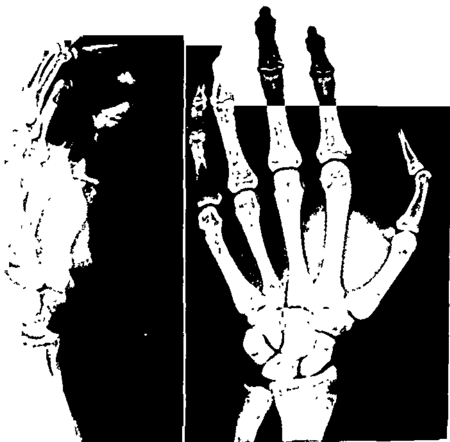


FIG. 2. There is satisfactory reduction, which was held in a position of flexion.

quent volar dislocation of the 2nd and the 3rd metacarpals.

Burk reports a case of dorsal dislocation of the 2nd and the 3rd metacarpals following a compressional force applied from the radial to the ulnar side when his patient's hand was pinned under a stone.

The exact mechanism of injury in the case reported is unknown. As noted previously, the patient was unable to describe the mechanism, and there were no skin abrasions to indicate that the wheel of the tractor had run over his hand.

Traction on the fingers coupled with pressure over the bases of the dislocated metacarpals is usually effective in reducing the dislocation if seen early. Late cases frequently are impossible to reduce by closed methods.

Bannes reports that he was unsuccessful in reducing a dorsal dislocation of the 2nd and the 3rd metacarpals, using traction on both fingers coupled with pressure over the bases. Of interest was the fact that traction on the 3rd metacarpal alone with pressure over the bases easily reduced the dislocation.

Bergasse and Guilmain were easily able to reduce the dorsal dislocation of the 3rd metacarpal; however, they had to resort to open reduction for the dislocation of the 2nd metacarpal.

Orrillard's case of vollar dislocation of the 2nd and the 3rd metacarpal was easily reduced and stable.

The position of stability for dorsal dislocation is extension. Many authors report recurrence of the dislocation following splinting in flexion. Santy reports what he felt to be a recurrence of dorsal dislocation of the 2nd and the 3rd metacarpal when splinted in flexion. Presumably, flexion is the position of stability for volar dislocation Most authors advise splinting for 2 to 4 weeks, depending on the stability at the time of reduction.

Whitson states that the reduction is usually more stable if the mechanism was one of indirect force.

If, following reduction, the dislocation is unstable, transversely placed Kirschner wires may be driven through the dislocated metacarpals into the adjacent normal metacarpal. Waugh and Yancey reported a case of dislocation of the 4th and the 5th metacarpals, first seen 3½ months following injury, satisfactorily treated this way.

Bunnell states that "reduction is necessary to restore muscle balance and proper mechanics in the hand." However, in most instances excellent functional results are reported in both those cases reduced with maintenance of the reduction, and those in whom reduction either was not done or in whom it was not maintained. Watson-Jones states, "There is relatively little functional disability even if such displacements are imperfectly corrected." Shorbe points this out, as do Waugh and Yancey, and Whitson. Therefore, careful evaluation is necessary for those patients presenting with old dislocations before open reduction is resorted to. However, every attempt should be made to obtain reduction in those cases seen early and in those patients who require excellent hand function.

BIBLIOGRAPHY

Bannes, F.: Zur Kasuistik der Luxatio carpometacarpea, Deutsche med. Wchnschr. 28: 608-610, 1902.

Berdach, Julius, and Herzog, Alois: Ein Fall von traumatischer, isolierter Luxation des Metacarpus indicis, Wien. klin. Wchnschr. 15:940-942, 1902.

Bergasse and Guilmain: Des luxations dorsales, Arch. méd. et pharm. mil. 62:284, 1913.

Biancheri, T. M.: Le lesioni traumatiche del carpo, Chir. org. movimento 4:347-392, 1920.

Blandin: Luxation incomplète du troisième métacarpien en haut, J. connaissances méd.-chir. 12:177-179, 1844.

Bourguet: Observations pour servir à l'histoire des luxations des doigts et de la main: Obs. VIII. Luxation incomplète du second métacarpien en avant sans division des téguments; réduction le sixième jour de l'accident, Rev. méd.-chir. 14:94-95, 1953.

Bunnell, Sterling: Surgery of the Hand, ed. 3, pp. 816-817, Philadelphia, Lippincott, 1956.

Burk: Ueber die Luxatio carpo-metacarpea, Beitr. klin. Chir. 30:525-545, 1901.
Buzby, B. F.: Palmar carpo-metacarpal dislocation of the fifth metacarpal, Ann. Surg. 100:555-557, 1934.
Grant, J. C.: A Method of Anatomy: Descriptive and Deductive, ed. 5, pp. 139-140, Baltimore, Williams & Wilkins, 1952.
Hamilton, F. H.: A Practical Treatise on Fractures and Dislocations, ed. 7, p. 794, Philadelphia, Lea, 1884.
Hammann: Über die Luxatio carpo-metacarpea, Deutsche Ztschr. Chir. 223:287-296, 1930.
Humbert, G.: Luxation du deuxième métacarpien en arrière, Union méd. 5:527-528, 1868.
Key, J. A., and Conwell, H. E.: The Management of Fractures, Dislocations and Sprains, ed. 5, p. 767, St. Louis, Mosby, 1951.
Lyman, C. B.: Backward dislocation of the second carpo-metacarpal articulation, Ann. Surg. 43:905-906, 1906.
Morris: Human Anatomy, ed. 10, p. 325, New York, Blakiston Division of McGraw-Hill, 1942.
Orrillard, A.: Des luxations de l'extrémité supérieure des quatre derniers métacarpiens dans leurs articulations avec le carpe, Gaz. hôp. 66:1085-1092, 1893.
Roux, Jules: Luxation des os du métacarpe dans leur articulation carpo-métacarpienne, Union méd., p. 224, 1848.
Santy: Luxation carpo-métacarpienne et tarso-métatarsienne, Lyon chir. 20:400-401, 1923.
Shorbe, H. B.: Carpometacarpal dislocations: report of a case, J. Bone & Joint Surg. 20: 454-457, 1938.
Watson-Jones, R.: Fractures and Joint Injuries, ed. 4, pp. 635-637, Baltimore, Williams & Wilkins, 1955.
Waugh, Richey L., and Yancey, Asa G.: Carpometacarpal dislocations (with particular reference to the simultaneous dislocation of the bases of the fourth and fifth metacarpals), J. Bone & Joint Surg. 30A:397-404, 1948.
Whitson, R. O.: Carpometacarpal dislocation: a case report *in* Clinical Orthopaedics, No. 6, pp. 189-195, Philadelphia, Lippincott, 1955.

Dislocation Carpo-Metacarpal Reporto de un Caso

Summario in Interlingua

Es reportate un caso de dislocation palmar del secunde e tertie metacarpales. Isto es le dece-prime caso de dislocation del secunde e tertie metacarpales reportate in le litteratura.

In le presente caso le exacte mechanismo non es cognoscite. Le reduction esseva effectuate sub un pro cento de procaina, infiltrate localmente, per traction al secunde e tertie digitos con pression directe super le bases del dislocate metacarpales. Le immobilisation esseva effectuate in un position de flexion. Illo esseva mantenite durante cinque septimanas. Le plus grande stabilitate post-reductional es assecurate in le position de extension in casos de dislocation dorsal e in le position de flexion in casos de dislocation palmar. *Le reportos publicate include* casos de trauma directe e de trauma indirecte como causa de dislocation carpo-metacarpal.

Reduction per methodos claudite deberea esser essayate in casos precoce. Usualmente illo succede e remane stabile. Casos a presentation tardive require possibilemente un reduction aperte con fixation al adjacente metacarpal normal effectuate per medio de filos de Kirschner in placiamento transverse. Le perdita functional que resulta del non-reduction o del reduction imperfecte de tal dislocationes es inconsiderabile. Per consequente, casos de presentation tardive deberea esser tractate per reduction aperte solmente post le plus caute reflexion.

24

The Physician and His Hospital

R. T. MCELVENNY, M.D.*

The cost of medical care has become a popular subject. Certain writers have seized upon our medical profession and have tried to have us enter into acrimonious debates through the medium of the popular press. These writers should define and delineate their statements. Medical expense or cost is that money paid to the physician for his services. Hospital cost is that money paid to a hospital for board, room, laundry and personal services. Laboratory cost is that money paid for ancillary services expressed by various tests and procedures. Drug cost is that money paid for various therapeutic aids administered to the patient. Hospital charges today usually include, in itemized form, the cost of ancillary services and drugs, but the charges for *nursing services are masked.*

Hospital costs run at high level, because hospitals are not hotels, not homes, not doctors' workshops, and not business establishments. To save life they require different construction and different attitudes from anything else. Their equipment must be varied and unusual, and though some of it is seldom used, it is essential when needed. A good hospital has this depth of reserve. This is its function. It is built to preserve and save life.

The time element has much to do with these costs It costs far more today per hour to treat a person. Money is demanded from the patient in greater amounts at shorter intervals; whether it gives more in purchasing power than that which was dribbled out over days and weeks formerly is debatable.

* Chicago, Ill.

Further, more people are going out the front door well than formerly. Also, their stay is shorter, and thus their total productiveness on the job in man-hour days is greater. This should be computed and deducted from the hospital expense. Insurancewise, it is not the unit cost that is so discouraging. It is the increasing number of units multiplied by the cost. Health or life is an intangible. Once either is lost, there is no substitution or sublimation for it. To preserve it or save it costs money and, as more ways are found to do this, the cost per hour increases. You do it, or you don't do it; it is an "all or none" proposition.

There are some problems of cost that should be mentioned. The money necessary to keep a hospital open for giving service to people must be obtained. To many of us some methods of obtaining this money seem to be somewhat odd. We do not question the amount of money needed. We question only the method of obtaining it.

For instance, hospitals usually load the charges for ancillary services and drugs instead of further increasing the room rate. This practice is deplorable because:

1. Patients who require extra services are forced to carry part of the room charge load of patients who do not require extensive ancillary services.

2. Most ancillary services, such as laboratory, roentgenology, pathology and anesthesia end in figures, pictures, slides or machines, all of which are inanimate and mean nothing until brought to life by a trained, skilled physician. The cost, plus a reason-

Burk: Ueber die Luxatio carpo-metacarpea, Beitr. klin. Chir. 30:525-545, 1901.
Buzby, B. F.: Palmar carpo-metacarpal dislocation of the fifth metacarpal, Ann. Surg. 100:555-557, 1934.
Grant, J. C.: A Method of Anatomy: Descriptive and Deductive, ed. 5, pp. 139-140, Baltimore, Williams & Wilkins, 1952.
Hamilton, F. H.: A Practical Treatise on Fractures and Dislocations, ed. 7, p. 794, Philadelphia, Lea, 1884.
Hammann: Über die Luxatio carpo-metacarpea, Deutsche Ztschr. Chir. 223:287-296, 1930.
Humbert, G.: Luxation du deuxième métacarpien en arrière, Union méd. 5:527-528, 1868.
Key, J. A., and Conwell, H. E.: The Management of Fractures, Dislocations and Sprains, ed. 5, p. 767, St. Louis, Mosby, 1951.
Lyman, C. B.: Backward dislocation of the second carpo-metacarpal articulation, Ann. Surg. 43:905-906, 1906.
Morris: Human Anatomy, ed. 10, p. 325, New York, Blakiston Division of McGraw-Hill, 1942.
Orrillard, A.: Des luxations de l'extrémité supérieure des quatre derniers métacarpiens dans leurs articulations avec le carpe, Gaz. hôp. 66:1085-1092, 1893.
Roux, Jules: Luxation des os du métacarpe dans leur articulation carpo-métacarpienne, Union méd., p. 224, 1848.
Santy: Luxation carpo-métacarpienne et tarso-métatarsienne, Lyon chir. 20:400-401, 1923.
Shorbe, H. B.: Carpometacarpal dislocations: report of a case, J. Bone & Joint Surg 20: 454-457, 1938.
Watson-Jones, R.: Fractures and Joint Injuries, ed. 4, pp. 635-637, Baltimore, Williams & Wilkins, 1955.
Waugh, Richey L., and Yancey, Asa G.: Carpometacarpal dislocations (with particular reference to the simultaneous dislocation of the bases of the fourth and fifth metacarpals), J. Bone & Joint Surg. 30A:397-404, 1948.
Whitson, R. O.: Carpometacarpal dislocation: a case report *in* Clinical Orthopaedics, No. 6, pp. 189-195, Philadelphia, Lippincott, 1955.

Dislocation Carpo-Metacarpal Reporto de un Caso

Summario in Interlingua

Es reportate un caso de dislocation palmar del secunde e tertie metacarpales. Isto es le dece-prime caso de dislocation del secunde e tertie metacarpales reportate in le litteratura.

In le presente caso le exacte mechanismo non es cognoscite. Le reduction esseva effectuate sub un pro cento de procaina, infiltrate localmente, per traction al secunde e tertie digitos con pression directe super le bases del dislocate metacarpales. Le immobilisation esseva effectuate in un position de flexion Illo esseva mantenite durante cinque septimanas. Le plus grande stabilitate post-reductional es assecurate in le position de extension in casos de dislocation dorsal e in le position de flexion in casos de dislocation palmar. Le reportos publicate include casos de trauma directe e de trauma indirecte como causa de dislocation carpometacarpal.

Reduction per methodos claudite deberea esser essayate in casos precoce. Usualmente illo succede e remane stabile. Casos a presentation tardive require possibilemente un reduction aperte con fixation al adjacente metacarpal normal effectuate per medio de filos de Kirschner in placiamento transverse. Le perdita functional que resulta del non-reduction o del reduction imperfecte de tal dislocationes es inconsiderabile. Per consequente, casos de presentation tardive deberea esser tractate per reduction aperte solmente post le plus caute reflexion.

24

The Physician and His Hospital

R. T. McELVENNY, M.D.*

The cost of medical care has become a popular subject. Certain writers have seized upon our medical profession and have tried to have us enter into acrimonious debates through the medium of the popular press. These writers should define and delineate their statements. Medical expense or cost is that money paid to the physician for his services. Hospital cost is that money paid to a hospital for board, room, laundry and personal services. Laboratory cost is that money paid for ancillary services expressed by various tests and procedures. Drug cost is that money paid for various therapeutic aids administered to the patient. Hospital charges today usually include, in itemized form, the cost of ancillary services and drugs, but the charges for *nursing services are masked.*

Hospital costs run at high level, because hospitals are not hotels, not homes, not doctors' workshops, and not business establishments. To save life they require different construction and different attitudes from anything else. Their equipment must be varied and unusual, and though some of it is seldom used, it is essential when needed. A good hospital has this depth of reserve. This is its function. It is built to preserve and save life.

The time element has much to do with these costs. It costs far more today per hour to treat a person. Money is demanded from the patient in greater amounts at shorter intervals; whether it gives more in purchasing power than that which was dribbled out over days and weeks formerly is debatable.

* Chicago, Ill.

Further, more people are going out the front door well than formerly. Also, their stay is shorter, and thus their total productiveness on the job in man-hour days is greater. This should be computed and deducted from the hospital expense. Insurancewise, it is not the unit cost that is so discouraging. It is the increasing number of units multiplied by the cost. Health or life is an intangible. Once either is lost, there is no substitution or sublimation for it. To preserve it or save it costs money and, as more ways are found to do this, the cost per hour increases. You do it, or you don't do it; it is an "all or none" proposition.

There are some problems of cost that should be mentioned. The money necessary to keep a hospital open for giving service to people must be obtained. To many of us some methods of obtaining this money seem to be somewhat odd. We do not question the amount of money needed. We question only the *method of obtaining it.*

For instance, hospitals usually load the charges for ancillary services and drugs instead of further increasing the *room* rate. This practice is deplorable because:

1. Patients who require extra services are forced to carry part of the room charge load of patients who do not require extensive ancillary services.

2. Most ancillary services, such as laboratory, roentgenology, pathology and anesthesia end in figures, pictures, slides or machines, all of which are inanimate and mean nothing until brought to life by a trained, skilled physician. The cost, plus a reason-

able overcharge for these inanimate objects, is easily determined and is the fair charge. Hospitals usually charge this, plus the interpretation of the physician. These services belong to medical personnel and are of value only in this sphere. This sad situation either must be changed or all must suffer in silence.

3. Free care by voluntary hospitals, given from running expenses, is becoming quite unusual in the present day. Various charitable funds, agencies and insurance plans have taken over the free bed to the point that, while it may be free to the patient, the hospital is reimbursed for the most part. The professional services provided by the physician in most cases are given with little or no charge.

Hospital management should be required to charge adequately for the services that they can render but to stay out of the sphere of the physician. Then the cost of ancillary services can be somewhat more realistic.

We have waited in vain for a popular article to state clearly that medical fees that the patient pays the physician have not even doubled in the past 15 years, while during *this time hospital costs have trebled or* quadrupled, as have drug costs.

In the author's territory, prior to 1945, the National Foundation for Infantile Paralysis paid the physician $75 for a major procedure. At the same time, this foundation paid the hospital $4.40 a day for the patient's care.

At the present time, this foundation pays the physician $100 for any major procedure or multiples of it done at one time for the same condition. The top fee is $175 for multiple operations, multiple cast changes and other care, even in a long-drawn-out procedure such as treatment of spinal curvature. The hospital now receives a basic remuneration from this foundation of approximately $21 a day. The physician's fee (and these fees are set by committees of physicians working with the foundation) has risen from $75 to $100 for a major procedure, while the hospital cost has risen from $4.40 a day to $21 a day.

Those who have written on the subject of hospital charges have failed to show the division of costs and services between those that are attributable to the lay management and those that come from the physician. Only by a comparison of these is a fair interpretation possible.

In the public's mind, cost to the patient for his physician, his hospital and his drugs are all grouped at the physician's level. This fact should be welcomed by the medical fraternity, but if we accept this responsibility, we have a covenant with the public to assume the responsibility actively. If we do so, we can maintain our many special privileges. To point this up, a few facts should be cited, namely:

1. The quality of a hospital staff determines hospital occupancy.

2. The physician originates the occupant of the hospital. If he originates the occupant, he eventually could control the source of hospital usage.

3. *If the physician controls this usage, he* can determine to a large extent where, when, *why and how a patient enters a hospital.* This has great bearing on hospital management, hospital service, hospital costs, hospital construction and hospital utilization.

4. The physician must learn that control of source, or the control of the money that determines source, must ultimately determine program. Our profession must participate in this problem of control if we are not to betray the public and allow them to prove to themselves how mediocre care can become when others control source, supply, facilities and the physician himself.

5. More and more the physician is allowing the hospital to become involved with expensive facilities for treating the sick and then utilizing these facilities for examination of the presumably well. This unnecessary utilization points up two facts:

A. Many people are going about their daily lives with conditions that, with early

diagnosis, might have been prevented or corrected.

B. Hospital space meant for preservation of life is utilized for care of people who are not sick.

These two factors tie in with rise or fall of hospital costs. It is evident that, only as medical expense rises slightly, can hospital costs fall markedly. The net profit to the consumer lies in a halt in the rise of hospital insurance costs through diminished unnecessary hospital utilization. The physician's services are present, whether or not the patient is in a hospital.

The public has the belief that diagnostic aids, known as tests, are the things on which a diagnosis or detection is made. These diagnostic aids mean nothing until evaluated by an able diagnostician. In one series I know of, laboratory examinations taken in mass turned up only 30 per cent of conditions unearthed. The diagnosticians, by careful and detailed histories and examinations, diagnosed the other 70 per cent. These tests are but tools of good medicine. It certainly is not necessary to build hospital beds to provide these tests.

A hospital is a place offering superior equipment for the use of a physician in treating the sick and the injured and is geared to this end. A doctor's office is geared to interviewing and, at times, treating the sick and the injured. Diagnostic aids are needed between the office and the hospital, and these can be erected at much less cost than are hospital beds. Such centers should be created in conjunction with any group of medical examiners skilled in diagnosis of physical and mental states. They should be established as extensions either of hospitals or of physicians' offices.

These establishments are geared and designed for the consumer and establish free communication between the patient and the physician. The patient sees that his physician, rather than the tests themselves, is of primary importance to diagnosis of his complaint.

Facilities of this nature would save many hospital beds so much more needed for definitive treatment of the sick, for research and for the training of youth.

One such facility is compiling statistics that are appalling in the frequency of pathology revealed in physical and mental surveys in presumably healthy individuals, 70 per cent of whom are under the age of 50. In most instances, the individual was unaware of his condition.

To attack this problem, we pose a few questions:

1. Has this patient ever been examined by a physician?
2. Would this newly discovered condition be found by any physician?
3. Do these facts indicate that physicians who concentrate on this work in a place designed for complete examination and diagnostic work-up unearth conditions and problems never before appreciated?

THE PHYSICIAN'S FEE

The quality of the physician and his hospital makes them deliver the same bill of goods to the indigent patient as to the one who can and does pay well. This quality of heart, head and hand must not betray us.

We know little of pay-for-service; in fact, many of our services are never paid for. We know mainly that we try to maintain and improve upon the things we do well. We do not fulfill desires of people; rather, we succor people in distress, without regard to race, creed, color or financial status. In no other realm but the church is service to people given so freely.

There is no such thing as "cheap" medical care, and people should be made aware of this fact. Good service is, of necessity, expensive service. The test of cost of any service is not the cost per unit, but the total cost of all units of service delivered. Therefore, as regards the sick and the injured, we hope to treat fewer and fewer of them for more and more money per unit. For the well, we

hope to examine more and more for less and less per unit.

Statistics to reveal the actual cost per unit of physicians' service should include all services delivered by physicians—to totally free and part-pay patients, as well as to full-pay patients.

THE PHYSICIAN IN RELATION TO CORPORATIONS

We speak of our cathedrals of learning as educational institutions, but they are mere repositories of accumulated knowledge. They produce educated people only if they are staffed by educators. An educator is a man who leads a mind forth. He stimulates it to gather knowledge, not for the sake of static parroting of what is known or thought to be known but to stimulate visions of things new, good, or useful. An educated man does not require a college degree; he requires only an endowment of capacity and the blessing of an educator. Then he produces.

Today, many people attend higher institutions of learning and acquire new tools, with which they explore new fields. Many give little thought or consideration to the tremendous importance of tradition and heritage.

The term "doctor" means teacher or guide. Historically, members of the professions have been the educated ones, the learned ones, the molders of society. The good brains of these groups have been beautiful, gentle and gifted; their predominant characteristics have been compassion and subtlety. They have served as advisers and referees through the ages.

Times may have changed. Technologic advances have been great, and some of them horrifying. The professions should take an active part in the applications of these advancements.

In a look at ourselves, certain limitations become evident

1. We lack standards of evaluation, statistically speaking

A. Often we cite too few cases.

B. We have not done enough statistical work to prove the things that we contend.

2. Our historical viewpoint of the physician sometimes fogs our perception of present-day medical needs. In times past, the patient first collapsed and then was carried away.

3. Preventive medicine at its present stage is concerned mainly with early detection. We examine the patient's body, explore his frustrations and his complexes and stir up the fire of his Id. Then we hustle him off to a foreign environment where he must make new adjustments and be introduced to new tensions and old loneliness, on the theory that this will cure his difficulties in his regular environment. What happens? He spends 2 weeks or 6 months and then returns to the old grind, with the old chains that bind him. He has not worked out a single problem in his home environment. He has only been fattened for the kill.

These factors have great bearing on the conduct of modern management, whether private or governmental. Management will spend any amount of money to reduce unit cost and eliminate "cripples" in the production line. Management spends a great deal recruiting and training men. If the question of saving these employees' most precious and irreplaceable element — the mind — arises, management will buy health resorts, duck blinds, hotels and even hospitals, but how little they spend on the essential ingredient —the physician!

Management must understand that a hospital should not serve as an escape. The physician, by dealing with the active problems, can teach the individual that he cannot escape and that he must work out his problems while maintaining his environment. Management must realize that peace of mind means rest in normal environment—a state far superior to rest, without peace of mind, in an abnormal environment.

Management must understand that recreation has a place but is not a cure, and some-

times not much help over a life span. Management must persuade the individual that he must either modify himself or his environment. Management must be shown that there is no such thing as "cheap" medical care.

Top management should concede that it needs a physician to sit in on top decisions regarding care of employes, and that the cost of this strategist in the most vital field of all—the individual—comes high. All business attempts to get larger and larger aggregates of capital in an attempt to cut down unit cost, without realizing that frequently the cost of the physician is offset by the great reduction in the casualty list.

Let us cite a few points concerning management:

1. A large business organization sends many of its key men on a flight which is lost. What then?

2. An executive starts for Chicago and ends in Louisville, a victim of amnesia. This man's company has a large medical setup, but the staff is working on the routine examination level.

3. A great leader, who anyone could see was only a shell, is allowed to guide his people at world-shaping conferences. What is the medical story on this?

4. The motor car manufacturers need top medical thinking. We need cars that are geared for our present roads and people—not geared for conditions that may exist 25 years from now. We need cars designed to preserve life and limb and, if this were done, thousands more car owners would be around to buy new models, and the medical and funeral expenses would go down surprisingly.

Why do situations such as these exist? In the first place, the physician has not taught management to think correctly about these problems, because the physician has not thought himself into the problem, and therefore has not become a teacher and a strategist for top management. Secondly, many times the physician in industry has taken too cheap a view of his own function.

In a corporation, as in many other sociologic phenomena, good things often start at the top and then filter down. To reach the masses, the enlistment of management is essential. Therefore, our main duty in industrial medicine is to determine the environment factors that enable a man to enjoy his job, his home and himself.

If we, as physicians, think ourselves into this problem of becoming advisers and strategists for management, we can, by our peculiar background, do much. When in top-level conferences, the physician must ask himself, "What will this do to the people involved in this decision?"

CONCLUSIONS

Physicians can help correct the lack of awareness, lack of courage and lack of vision that characterize many of our modern tools of living.

Also, they can cut down phony escapes into hospitals, nursing homes, sanatoria and watering places.

The physician must become more active in these spheres and he must interject his thinking more realistically into the planning of management, be it private or governmental in nature.

25

The Expert Medical Witness

PATRICK C. DORAN, M.D.*

Industry makes great efforts and expends large funds to improve public relations. Private utilities, the huge power, light and fuel companies, are especially concerned. As relative monopolies, they realize that the power to tax and to regulate, so closely allied with public opinion, may suppress or eliminate them. A fairly accurate index of public favor or dislike is available for tabulation in the press and in all methods of publication. Public relations departments analyze and grade the reactions. On this basis public disfavor of the medical profession is greater than of the private utilities. As an individual the doctor is beloved. As a group the medical profession is distrusted.

Public relations have been considered seriously by profound members of the profession. The address of the chairman of the American Academy of Orthopaedic Surgeons, Dr. Guy Caldwell, was devoted to this subject. The American Medical Association nationally and many of the state and the local societies are becoming conscious of the need to improve this phase of medical practice. A facet of public relations to which all doctors must be exposed is the medicolegal aspect. This arises mostly out of either of two forces—trauma or crime.

The average physician will not concern himself too much with crime, as this belongs to forensic medicine. In this, a traumatic age, one rarely may isolate medical practice from the medicolegal aspects of trauma.

* Akron, Ohio

TOTAL ACCIDENTAL INJURIES, 1953*

1. Motor vehicle	1,250,000
2. Public nonwork nonmotor vehicle	2,000,000
3. Industry	2,000,000
4. Home	4,350,000
	9,600,000

* National Safety Council: Accident Facts, 1954.

CLASS 1. MOTOR VEHICLE

A million and a quarter motor-vehicle injuries reported. Perhaps there were additional ones; especially so in some states that do not require or enforce the 24-hour injury report law. Since most of these occurred in the more populous areas, where the report laws and enforced insurance coverage are the rule, a goodly proportion of the number required investigation, adjudication or litigation. A fair estimate puts about 10 per cent, or roughly 100,000, involving the filing of a law suit, if not formal trial.

CLASS 2. PUBLIC NONWORK NONMOTOR VEHICLE

Two million injuries represent, possibly, a lower rate of medicolegal union. Nevertheless, the mode of injury in this category frequently connotes insurance coverage. Injuries occurring in merchandising establishments, elevator doors, revolving doors, slippery floors, escalators and such, where liability was established demanded medical professional care and its associated medicolegal attention.

CLASS 3. INJURIES IN INDUSTRY

Another 2 *million injuries* indicate complete professional attention—not necessarily doctors of medicine. To be valid, an industrial commission or an insurance work claim must have a practitioner's signature. Most of these cases may have been trivial, but 100,000 or more—based upon a state average—were serious. They required disability analysis, litigation and/or formal medicolegal attention.

CLASS 4. HOME ACCIDENTAL INJURIES

A number almost equal to the other 3 offers the least opportunity for medicolegal participation. If liability insurance or comprehensive insurance was involved, nevertheless the occasion for controversy arose and the inevitable association, neutral examination and possible litigation.

Increasing numbers of injury claims are processed each year. Many disability reports, analyses and court trials are involved In the moderate-sized and the large-sized communities and municipalities many patients who survived after serious traumatic injuries were the orthopaedist's problems. It is reasonably *certain* that the care was excellent. However, one is not sure that the average orthopaedist has represented himself and his profession as excellently to the public and in court. Specifically, what kind of expert medical witness does an orthopaedist make?

There are many excellent textbooks on medicolegal medicine describing the "doctor in court" and his associating with the law. The subject is quite alive in the current medical literature. McBride's, Schneider's and Eaton's *instructional course lectures* on this subject at the Academy Meetings are crowded. The A M. A. national journal and all the state and the county publications show increasing interest in this phase of medical practice. No known assay of public reaction to this phase of public relations is recorded.

Three classifications of witnessing conditions are cited in the state of Ohio: (1) witness of fact; (2) professional medical witness; (3) medical expert witness.

As is true of any other citizen, a doctor may be subject to subpeona for witness of facts, professional or otherwise. If he was at the scene when a crime or an accident occurred, obviously he must be available for testimony. This fact has not been sufficiently impressed upon medical students and on house doctors whose contact with the patients and their records may be needed in court *at any time*. It should be reiterated that what is said or written about patients may receive headline publicity, or it may have to be repeated under oath in court. More emphasis should be placed on the need for the accuracy of medical records and for their sanctity.

A professional witness is recognized as the attending physician whose records and testimony are needed to outline the patient's case by description of care, disability, expense and the like. He may or may not necessarily be an expert.

The third category—and the one most applicable—is the expert medical witness, defined as one whose qualifications, experience and character or integrity make him invaluable, possibly indispensable, in the matter of disability analysis in compensation work or testimony. Apparently, the courts make no issue of his attributes. Credentials required by most accredited hospitals for active staff appointment and by certifying bodies in medical practice and/or their equivalent are the accepted criteria.

Experience as a factor is difficult to define. It is that which the younger man thinks he *has but an older man knows he has not*. To quote a medical friend, "Experience certainly is not hereditary." Advice to the younger men and the beginners in court work may be summed up in a saying attributed to Davy Crockett, "Better get a lot of fur on your tail before they start shooting at you."

It was felt that doctors of medicine gen-

erally and orthopaedic surgeons specifically were of high integrity and good character. Snatches of social conversation with various groups indicated that this was not so. Therefore, an attempt was made on a local community basis to obtain an answer to certain questions which will be outlined. It was done in the mood of the philosopher, not in the role of the prosecutor. Interviewed at length were judges, industrial commission referees, adjusters, trial lawyers (both reputed plaintiff and defending types), court reporters and jurors.

QUESTIONS ASKED

Question 1. Do doctors (orthopaedists) impress you as wholly honest and sincere in their court testimony and in the medicolegal relations?

Question 2. What is your reaction when diametrically opposed testimony is offered by opposing experts (plaintiff's expert vs. defendant's expert)?

Question 3. What qualities and/or characteristics of the experts are most effective?

Question 4. Could you suggest any improvements?

Judges Interviewed

The 4 judges interviewed were common pleas or appellate jurists of 25 or more years' judicial experience. The common pleas court judges hear many personal injury trial cases, and the appeal court judge had and does have extensive judicial experience with this form of litigation.

Their answers to the questions were:

Question 1. Three, Yes. One, No.

Question 2. Two, "Differences are logical and honest," and 1 of them continued, "Take the United States Supreme Court, for instance; rarely is there a unanimous decision from this body." Two, "Doctors can be hired for any form of testimony "

Question 3. All 4 judges remarked, "Simplicity of language." Three mentioned an attitude of humility ("I could be wrong") Three said, "Effective speaking voice." All 4 said that the integrity, the reputation and the known character of the particular medical witness played a most important role.

Industrial Referees and Their Staffs (4 Individuals)

A word of explanation about the industrial referees and the hearings and the rehearings. These procedures are held in circumstances and in a calmer atmosphere than is the trial by jury. An expert is sworn, but the procedure is less formal. Rarely are more than 5 or 6 people present. All are seated. "Off the record" talks are held frequently. The hypothetic question comprises the nucleus of the situation. The pro or con role of the expert is to affirm or deny, principally the causative relationship of the hypothesis to the alleged disability.

The referees and their staffs hear much testimony from all varieties of medical expert witnesses. They answered the 4 questions as follows:

Question 1. Two, Yes. Two, No.

Question 2. "Doctors may honestly disagree. Medicine is an inexact science." Two said, "Doctors are for hire."

Question 3. Four agreed on 1 item, namely, that the reputation and the character of the expert witness comprised the most important factor.

Question 4. As to the improvement, they suggested that better records could be kept, especially as to what appeared to be a rather trifling matter of inaccuracies: distinction between right and left, for instance; or the identity of the finger or the toe involved.

Attorneys Interviewed

Twelve attorneys interviewed—all with extensive court experience, both in defense and plaintiff cases, answered:

Question 1. Eight, Yes. Four, No.

Question 2. Eight said essentially, "I can find a doctor to testify to anything needed." Four said, "Doctors are honest and not for hire."

Question 3. The attorneys cited again the character, the integrity and the reputation of

the medical expert witness. To quote some of their remarks: "An expert medical witness is one whose opinion I would accept on either side." "He doesn't need to be under oath." He settles the case out of court, etc."

Question 4. As to suggestions and to improvements, they all emphasized the need for pre-trial conferences with the doctors; the need for more prompt submission of reports and greater accuracy as to dates and other trivia not usually considered to be too important by the doctors.

Adjusters Interviewed

Four adjusters answered as follows:

Question 1. One, Yes. Three, No.

Question 2. The patient's expert—if he is one—is going "to slant his testimony in favor of his patient." Two said, "Most of them are honest, but naturally they are swayed by *the side that is paying them.*"

Question 3. All 4 agreed on age, experience and reputation.

Question 4. Conferences with the attorneys and the adjusters about the facts of the case. More prompt submission of reports; more accurate estimate of disability figures.

Court Reporters

Court reporters, two in number, were of no help. Both said that they were too busy taking down notes to analyze. However, both said that they wished that the doctor would talk louder and slower.

Jurors Interviewed

The jurors interviewed—with great difficulty—were asked one question only:

Question 1. The cases selected were those in which there was a wide difference of opinion between the attending doctor and the defense expert. Nevertheless, all felt that both doctors were honest. Such variations were explainable on the basis of logical difference of opinion One got the impression that the jurors saw the facts with the eyes of the attorney who impressed them most. A doctor's wife, who was a juror, said, "My husband differs and argues with his associates all the time. I suppose he would do the same in court."

CONCLUSION AND SUMMARY

On the whole, the reaction of various levels of the public to the medical professional expert witness (orthopaedists) was found to be quite favorable in a small sampling taken on a local community basis. The objectivity of most of the factors involved probably explains this fact. Cited most frequently as the *most important attribute to be brought to* court or to back a report was the personal reputation of a doctor. Of all the groups interviewed, the least impressed by the doctor's integrity were the industrial commission referees. At times, the fantastic connections made by testimony regarding the aggravating factors and the causes of some well-known disease of spontaneous onset probably were responsible for their derogatory attitude.

BIBLIOGRAPHY

Am. Coll. Surgeons: The hospital medical library, 1947.

Gonzales, T. A.: The needs for improvement in medicolegal investigations, New England J. Med. 215:385-388, 1936.

Magnuson, P. B.: The doctor as a witness before industrial commissions, n.d.

O'Dunne, E.: The doctor as a witness, M. Clin North America 25:303-311, 1941.

Regan, L. J.: Important aspects of legal medicine, Ann. West. Med. & Surg. 3:162-164, 1949.

Stone, R. A.: Medical testimony, Minnesota Med. 24:532, 1941.

Sulzberger, M., Jr.: What every physician should know about cross-examination, Bull. New York Acad Med. 18:665-692, 1942.

Trostler, I. S.: The physician as a witness, Illinois M. J. 104:189-193, 1953.

Winston, Smith Hubert, ed.: Scientific Proof and Relations of Law and Medicine, Urbana, Univ. Illinois, 1946.

Le Experte Testificator Medical

Summario in Interlingua

Commentos es presentate super le relationes public del profession medical. Es sublineate le facto que le majoritate del medicos associa "relationes public" con activitates medico-legal. Se signala le crescente incidentia de vulneres accidental. Es presentate estimationes del aspectos medico-legal del annual total de 10.000.000 casos de vulnere. Le varie rolos del medico como testificator es describite. Es sublineate in isto le qualificationes del testificante experto medical.

Esseva executate un scrutinio inter le gruppos e le institutiones que se trova professionalmente in intimissime association con medicos in le campo del activitates medico-legal. Judices, arbitros de Commissiones Industrial, adjustatores, advocatos, membros del personal de corte, e juratos esseva interrogate in re lor experientias e/o opiniones concernente *le qualificationes e le integritate de medicos in* le rolo de testificatores. Le responsas e reactiones esseva registrate.

De accordo con le expectationes, le responsas al quatro questiones usate variava considerabilemente. Certe responsas es citate in forma directe. Le majoritate del opiniones esseva favorabile. Il pare que on recognosce que le practicantes de un scientia inexacte ha le derecto de differer honestemente in lor opiniones.

SECTION III

Motorist Injuries and Motorist Safety

REDUCTION OF INJURIES (CRASH-IMPACT ENGINEERING)

(Part 2)

GUEST EDITOR: JACOB KULOWSKI, M.D.
Saint Joseph, Missouri

1

Introduction to Supplementary Safety: The Crash-Impact Engineering Point of View

JACOB KULOWSKI, M.D.

This symposium, written primarily for clinicians, is being presented in a sort of retrograde fashion; that is, from the resultant crash injuries through etiology and ultimately to the prevention of the accidents themselves. Part 2 will deal with etiologic factors in the physical sense of the word, or the so-called crash-impact engineering point of view, as it relates to crash injuries per se. The real causes of the accidents will be discussed in Part 3, to be published in the 1957 Spring Issue of *Clinical Orthopaedics*. It will become more and more evident to the reader that the problems involved in motoring injuries and motoring safety include a variety of psychosomatic and mechanical factors which tend to blur into one another as one delves deeper into its multifaceted aspects and challenges. This truth cannot dim the importance—human nature being what it is—of a discipline which aims at the reduction, prevention and/or moderation of injuries under crash decelerative conditions and/or upsets.

This is the essence of crash-impact engineering and design in respect to automobile accidental injuries and deaths. Clear cut as it is, the rapidly advancing knowledge regarding the problems of motoring safety demands some clarification of terms which might be confusing, especially when used out of their own particular contexts.

Automotive safety engineering has dealt with the traditional aspects of performance *reliability of the motor vehicle in question —for all practical purposes, the automobile driven by nonprofessional operators, but not excluding commercial carriers.* The current record of safety performance of modern automobiles hardly needs to be mentioned at this time; however, it will be discussed in detail in Part 3. This is done in order to distinguish that kind of safety engineering from what is termed automotive crash-impact safety engineering and design which forms the basis of this section of our symposium.

The term *human engineering* is comming to the forefront of motoring safety discussions and should not be confused with what has been defined above. Actually, human engineering deals with accident prevention rather than with reduction of injuries under crash conditions. It seeks to reconcile human psychomotor limitations with automotive engineering and design. In other words, human engineering seeks to render operation of the motor vehicle as foolproof as possible from the standpoint of safety. Human engineering assumes that automotive engineering and design can be brought within the limitations and the capabilities of the people who are expected to operate them safely as well as efficiently. Thus, human engineering will be discussed more fully in the next issue of the symposium.

The medical profession (clinical medicine

and surgery) owes a great debt of gratitude to crash-impact engineering because, prior to its advent, clinicians had paid little attention to the causes of the mechanical injuries received in crashes of motor vehicles. Hugh DeHaven was the first to emphasize the important relationships which existed between results of impacts and the impact areas. Thus, basic factors of mechanical causes (pressures) of these injuries became clarified and became the facts on which prophylaxis, in the physical sense, could be formulated. Since then, as will be shown in Part 3 of this symposium and the ever-increasing medical articles which are appearing in the journals, this deplorable situation is being remedied. Doctors everywhere should co-operate with the field studies currently under way and sponsored by the Cornell (Automotive) Crash Injury Research. In addition, medical groups should become more aware of what is being done in this field and offer their opinions and approval or disapproval of methods and approaches to such human salvage from automotive crashes. Finally, doctors should give scientific and financial support to the kind of work along these lines which they approve of. So much of what is being done begs for corroborative study and experimentation. This area of motoring safety happens to be the only one that is subject to experimental reproduction and study.

Be that as it may, valid medical data, both living morbidity and autopsy pathology, are essential to the success and the progress of crash-impact studies. Oddly enough, in this highly specialized field of scientific endeavor, careful clinical observations often assume a directional role in this respect no less than a supplementation of what experimenters disclose. The common sense and the intrinsic values of clinical studies and observations cannot be overestimated. However, the gap caused by lack of valid medical data between engineering and medicine is being bridged rapidly and positively. It is hoped that the final closure of this gap will eliminate muddled thinking and pitfalls to truthful conclusions in regard to the reduction of crash injuries. In order to attain this desirable goal, medicine will have to reach out from its narrow vertical specialty and broaden horizontally into domains closely related to the medical profession. The question is, "Will medicine finally awaken to its opportunities and responsibilities in the parallel field of ground traffic as it has in regard to flight transport?" The signs are that the answer will be for continued medical progress on an epidemiologic scale rather than in the strictly limited field of clinical practice. Truly, this is an area of interest where specialists become generalists and generalists become specialists without ever becoming aware of that fascinating transformation in either direction.

Another reason why crash-impact studies must be allowed to reach natural conclusions and results is derived from the fact that with the perfection of crashproofing, man's own inherent faults in motoring accidents will again come to the attention of the individual and the public.

In the meantime, the automobile—the chief counterpart of the human and inhuman relationships of accidental injuries—is receiving the bleakest kind of scrutiny and criticism from many sources of inquiry. This unilateral reaction is understandable to a great extent and will continue to be the case until automotive safety engineering and design will have reached an unimpeachable peak of achievement. If progress in human engineering has not kept pace with that, then man's limitations and incapabilities will continue to cause accidents, injuries and deaths; unless, also, progress has been made in adequate medical standards of driver licensure. This cycle of events epitomizes what is meant by the blurring of psychosomatic-mechanical elements into one great design involving motoring safety and motoring injuries.

Manufacturers should remember that even the ultimate in automation sometimes magnifies man's physiologic and psychic faults;

i.e., the easier it becomes to drive a car, the greater is the probability that the driver will lose attention and the more violent will be his reaction — extensor — to sudden jars, sounds and dislocations. Will increasing safety and human engineering and design have such trends in driver behavior?

Briefly expressed, crash-impact engineering and design may be stated to be an equation between human tolerances and decelerative forces (mechanical variables). From that standpoint, the first line of defense becomes the limits of factors of safety of the human body. The second line of defense is the structure which intervenes between riders and forces developed in the external automotive environment. However, this must be interpreted from two standpoints: the unrestrained occupant and the safety-suspended human body. For the former, forces mean for the most part the applied variety (impulsive and crushing). All kinds of injuries are likely to result from this kind of force intervention. On the other hand, the properly suspended body will experience inertial forces under decelerative conditions. Since crash decelerations are very brief, this means that stresses will be concentrated upon supporting structures of the body (organ suspensions as well as other ligamentous and bony structures). In other words, under such conditions, when the limit of tolerance has been reached, internal injuries are to be expected, rather than external ones, such as result from applied forces.

Forcible ejection from the crash vehicle is a special case in which occupants are exposed to double jeopardy because their chances of serious injury and death are intensified. In this case all kinds of forces may operate upon the body both internally and externally.

Both deformation of vehicular structure and injury represent an absorption of crash energy beyond the capacity of the part to take it up without external evidence of strain or complete dissolution. Both manifestations are analogous events in separate domains of action and reaction. Thus, DeHaven simply expressed the basic relationship between injury and impact or pressure design. This relationship dominates all crash-impact thinking and methods of study. Prior to that idea, it was believed quite generally that injuries were haphazard events and that the limits of safety factors were entirely subordinated to the magnitude of the crash forces involved in the various accidents. Stated another way, this would mean that the former were intrinsically incapable of coping with the inexorably large forces which were governed by immutable and inviolable physical laws of mechanics.

The magnitude of the forces generated in the external environment by crashes is not too difficult to calculate. The same cannot be said for what happens in the internal automotive environment to riders during accidents. Be that as it may, evidence is accumulating all the time to indicate and to prove that design factors play a significant part in the production or the prevention of injuries. That is not to say that force magnitude does not play a decisive role in this, but rather that impact areas in themselves determine to a large extent what part of the body is to be injured, the degree of injury and the frequency of such injuries under comparable accident situations. For this reason seating in crash vehicles has clinical significance. To some extent, this determines the relationship between riders and impact areas in their immediate vicinity, provided that gross body dislocations do not alter this relationship too greatly.

Both the human variables (regarding tolerances to forces) and the mechanical variables involved in crashes determine the gross differences in the results as seen clinically; that is, the uninjured, the injured and the dead This distinction is easier said than visualized but exists nevertheless. The first subgroup is a constant reminder that interior automotive design is not entirely lethal or injurious to all occupants of crash vehicles. It serves to temper overhasty conclusions along this line.

CONCLUSIONS

A potent approach to motoring safety has emerged in the form of crash-impact engineering points of view. The causal relationship between the impact area and the injury has been established. Medical data and studies have been of real assistance to this end. However, more of such effort is needed. If the causes of crash injuries are established it seems reasonable to assume that predetermined modifications of automotive design should reduce or moderate injuries received during crashes. That is the premise upon which crash-impact engineering rests. Equally important is the realization that crash-impact engineering, like all other facets of this problem, is only another link in the chain of endeavor to bring motoring safety under better control. The crash-impact approach to motoring safety epitomizes the epidemiologic method; i.e., studying the interactions between host (rider), agent (vehicle) and environment (highway).

2

History of Auto Crash Injury Research: Police Point of View

FRANK A. JESSUP* AND ELMER C. PAUL†

Police officers, next to doctors, have been viewing the growing problem of automobile and other motor vehicular accidents and resultant injuries with increasing alarm, especially from the standpoint of the injuries themselves. It seemed to be only a short step from the human effects of crash forces to their possible causes or relationships to environmental factors such as automotive design and engineering in both the internal and the external automotive areas. Yet, it was not until January 1, 1949, that the first Auto Crash Injury Research had been completed by an officer of the Indiana State Police department. By that is meant a report comprehensive enough to have taken into consideration the possible relationships between effects of impacts and the impact areas themselves, in so far as it was possible to reconstruct the sequence of events which had taken place during the crash and/or the upset. Actually, the fundamental idea embracing such comprehensive police reporting of accidents was conceived some years previously by the observations of individual police officers who had been noting for some years the repetitive nature of the bodily injuries. Thus, the germ of the auto crash-impact interrelationships was born and developed subsequently to its present preeminence.

The core of Auto Crash Injury Research (Indiana) was derived from the question, "Since, or if, the nature of crash injuries is repetitive or predictable in type and bodily distribution, is it not possible to remedy this situation by what might be called crash-proofing automotive engineering and design as already had been underway in regard to aircraft design?" Obviously, the answer seemed to be in the affirmative. The first obstacle to overcome was the lack of valid engineering and medical data on which to base recommendations to automobile manufacturers in respect to improved automotive safety design and engineering—the kind of automotive design that should prevent or moderate crash injuries. Its initiation and ultimate accomplishment demanded new technics of crash reporting, in the first place, before adequate data could be accumulated and presented to the manufacturers of automotive ground vehicles. One of us (Elmer Paul) thought out the details of such a program in Indiana, convinced others of its feasibility and obtained permission to proceed with it. His plan contained no procedure for formulating or outlining an essential data reporting form. The final form included accident facts, crash deformation facts, medical data and photographic-on-the spot proofs. These three phases of the report, namely, vehicle report, medical report and pictures, made up a complete and detailed case history of the course of events—a unique achievement in the annals of com-

* Superintendent, Indiana State Police.
† Sergeant, Indiana State Police.

prehensive police observation and reporting with respect to automobile accidents and the results of these accidents including both vehicular structure and human beings.

In the first place it was necessary, for the pilot study, to limit the study to fatal accidents only and to those which had occurred in a selected area or county. From this, the program was expanded in 1951 to a statewide program of special (crash-impact) fatal accident study of rural Indiana. As was to be expected this kind of study soon brought into focus related data never before obtainable or available regarding automobile accidents. In fact, now for the first time there was statistical evidence to support our basic hypothesis of the repetitive nature of crash injuries to riders. This was especially notable with regard to such factors as opening of doors (forcibly ejecting occupants, thus increasing their chances for fatal injuries); windshield, dashboard and steering control impacts, etc. These categories of investigation came to light with such clarity that they left little doubt regarding their validity. Moreover, as future data accumulated, the fundamental nature of crash injuries was further substantiated along these lines. Soon, the evidence attracted other persons and groups in similar approaches to the study of automobile accidents, injuries and deaths. All of this activity has accelerated crash-impact thinking and action and has served to broaden the scope of the work. At the present time, the value and the significance *of an integrated form of police reporting is* being recognized increasingly throughout the country.

Among these who are making these studies may be mentioned medical groups and individuals, research laboratories and, more recently, the manufacturers themselves. It was fortunate that such a well-informed and well-organized group as the Automotive (Cornell) Crash Injury Research workers came into this field after their significant pioneering in the field of safety aircraft engineering design to reduce injuries and fatalities in this form of transportation. The latter did much to improve and expand the reportial forms we had been using in our own Indiana research. From this beginning, other states were induced to participate in the comprehensive programs of fact gathering, ultimately to be processed and evaluated by Cornell's staff of experts. The mighty oak signifying the completion of auto crash injury research has yet to grow to full fruition.

One point which needs to be emphasized, since the public is not aware of it, is the fact that, from the beginning, automobile manufacturers were aware of and approved such a fact-gathering of traffic accidents. Similarly, all others interested in the problem of supplementary safety were given information as it became available. From this dissemination of data there has been a *greatly* accelerated interest in motoring safety from all angles of approach, including accident prevention as well as aids to recovery after accidents, and the final rehabilitation of the victims. This stimulation of interest and activity all along the line has been one of the most outstanding examples of what valid thinking, prompt action and reliable data are capable of doing in a democratic environment to attack a vital challenge to our public safety and health.

But the area of specific accomplishment from crash-injury research is chiefly in the so-called crash-impact engineering field; a field concerned primarily with the reduction, *the prevention and/or the moderation* of injuries under crash and/or upset conditions of deceleration. Already, things that appeared to be remote only a few years ago are appearing, as a routine matter, in automotive engineering design such as padded dashes, changes in steering controls, safety belts, as well as efforts at accident prevention through the advertising of manufacturers. This open affirmation of what is going on in this field is most encouraging to everybody concerned.

It should be remembered that crash-impact engineering is still in its infancy, but

already great strides have been made to give better protection to occupants of automobiles and other ground motor vehicles. A most valuable tool—the accident record now utilized—has been added to our armamentarium with which to bring automobile accidents, injuries and deaths under control. Data extracted from these records, when they become numerically adequate, will have given much food for thought and action to many persons and groups. Undoubtedly, then, the Auto Crash Injury Research (Indiana) will have reached its ultimate goal, that is, "to reduce the severity of injury to motor vehicle occupants when impact occurs."

SPECIAL COMMENT BY A POLICE ADMINISTRATOR (F.A.J.)

The police administrator, in his role of traffic control officer, is aware of his responsibility to maintain a safe, efficient and unobstructed flow of ground transport of people and goods. To this end, he must plan, in so far as is possible, to employ new methods of approach as they appear, even though such decisions entail further work and effort on the part of his already overburdened officers. A primary aim is to prevent accidents Since all kinds of approach serve toward that end, because of their educational values for the public, even the reduction of injuries can be interpreted in the light of accident prevention tools, but it also stands on its own merits as an autonomous facet in the fight for survival. Others are urged to participate in this program of reduction of injuries. It is not complicated. It is simply an extension of former police methods of accident investigation and reporting. Careful execution of this kind of reporting does furnish the automotive industry, police agencies, safety workers, clinicians and motor vehicle users with previously unknown specific facts concerning crashes and their chances for survival.

EDITORIAL NOTE (J.K.). The traffic safety worker often finds himself in a lonely spot. This is especially true of the conscientious police officer who tries to do what he can to stem the tide of rider injuries and deaths which threaten to engulf more and more (already on the second million in this country) of us in the blood bath of accidental crashes. Clinicians will do a real service to their fellow men by giving positive and vigorous support to their local police agencies who are trying to make our motoring safer.

No single person in traffic safety, from the standpoint of crash-impact engineering, deserves more credit (unless it be Hugh DeHaven) than modest Sergeant Elmer C. Paul.

It is to be remembered that no single approach to motoring injuries or safety can solve its manifold problems; nor can there be the ultimate accomplishment of integrating our efforts to the same end. The plain fact is this: that every single method of study and approach must be pursued to its own particular end; that is, as an isolated and autonomous human effort to engender a proper social and human attitude in drivers of motor vehicles.

From this angle, it is evident that Sergeant Paul's achievement has been a brave dream and a practical one indeed.

3

The Historical Development of the Crash-Impact Engineering Point of View*

A. HOWARD HASBROOK†

Protection from the injurious effects of "crash impact" originated when man, during the dawn of history, first fashioned a raw-hide shield to protect himself from the clubs, the spears and the arrows of his enemies. From that time until only recently, engineering consideration for designed protection from accidental injury or death was confined almost exclusively to the field of warfare.

The Vikings, for example, used protective head gear and shields; shields, which, when not carried individually, were placed on the sides of their boats for protection during shore attacks. Again, in later history, metal helmets, chain mail and shields were used by King Arthur's Knights of the Round Table for protection against injury in combat. Later, during the invasion and the conquest of Mexico, metal breastplates and helmets were worn by the Spanish conquistadors for protection from "crash impacts" of sword points, bullets, spears and arrows.

However, during periods of the Revolutionary War and the Civil War the need for individual personal protection was largely lost sight of; probably due to the need for operational freedom and mobility of the foot soldiers. However, the relatively low power and inaccuracy of the rifles and the pistols used at that time kept the casualty rate at a "reasonable" figure.

Moreover, up to and including the Civil War, the majority of deaths were due to disease. In relation to the catastrophic effects of the plagues and the epidemics that raged throughout the continents of the world, serious injury or loss of life resulting from the use of transportation media available during these years was of minor extent and importance.

However, many injuries and deaths were caused by persons being thrown from horses, chariots, wagons, carriages and even early steam trains. But, most of these incidents were looked upon by a superstitious populace as being the result of "bad luck" or acts of God. This reliance on "luck" apparently stifled any development of safety engineering or design for the protection of human life in accidents.

In addition, transportation media, and in particular the horse, did not lend themselves too well to designing for protection from injury in accidents. It is doubtful, for example, that anyone would have seriously considered

* This brilliant account of the history of the crash-impact engineering point of view, by the Director of Cornell University's Aviation Crash Injury Research, who has brought this phase of safety engineering into prominence in the air transport industry, here and abroad, shows how tortuous has been the route toward achieving reduction of personal injuries under crash and upset decelerative conditions. Much of the early progress was due to the persistence and the clear thinking of Hugh DeHaven, now retired. Only a limited number of people have been aware of this man's single purpose in life. Now, it is hoped, he will receive the honors and the credit that he has earned and so richly deserves. [Ed.]

†New York City.

tying himself to a saddle on a horse for protection against being thrown off, for it was essential, safetywise, to be able to throw one's self clear if the horse should fall accidentally.

Development and use of the gasoline engine in motor cars provided transport at higher speeds and thus created both a greater exposure to accidents (because of a lack of reflex training) and also a greater exposure to severe and fatal injury, due to the velocity of impact.

Also, during the era of the bicycle and the "horseless carriage" some imaginative and courageous men were endeavoring to learn the secret of flight. Lilienthal is one of the best known who succeeded in early times in flying in a heavier-than-air machine—a glider. The first to fly a powered heavier-than-air machine, of course, were the Wright brothers; those who followed in the footsteps of the Wrights developed flying machines that were capable of greater and greater speeds—speeds which eventually exceeded that of the fastest motor car.

Later, aircraft increased the exposure to injury in accidents by virtue of their higher speeds; the force of gravity acting on an airplane falling from a great height also tended to increase the force of impact.

Exposure to serious injury and death was also intensified in early aircraft because the occupants, in most cases, were located in the forward and most vulnerable end of the vehicle. Since these early aircraft were fragile and extremely liable to accidents, due to inadequate control and aerodynamic design, injury and death rates were high. Despite the obvious need for "tying in" the occupant, it was not until just prior to World War I that safety belts were first installed in aircraft, and soon became standard equipment in military aircraft. The need for tying the occupant to his vehicle was demonstrated vividly when Lieutenant Towers, who later became an admiral in the Navy, lost control of his aircraft and was thrown out of his seat in mid-air. Fortunately, he was able to grasp a portion of the airplane and thus was able to hang on until it crashed; he was seriously injured. The other occupant was reported to have been fatally injured.

However, installation and use of safety belts was based only on the need for restraining the occupants from falling out of the aircraft in turbulent air or during acrobatic maneuvers. Little or no consideration was given, up to that time, to the possibility that the safety belt might protect occupants from injury or death in a crash. In fact, many pilots would unfasten their safety belts if they knew they were going to crash because they feared that the safety belt might cut them in half. Many pilots felt that they had a better chance of surviving if they were thrown clear of the aircraft. Since postcrash fires were quite prevalent, it is possible that some did survive by being thrown clear.

A paradoxical point in the study of the development of crash-injury concepts was that the human body had been erroneously considered as a rather fragile object, and that protection from injury or death could be obtained only by preventing injury-producing accidents. Thus, accident prevention became the prime consideration in safety thinking and action.

However, the idea that the opposite was the case evolved in the mind of a young flying cadet who was involved in a mid-air collision in Canada during the latter part of World War I. In the resulting crash of two aircraft, this cadet, an American named Hugh DeHaven, was the only survivor. During the following months in the hospital, DeHaven kept asking himself "Why did I survive when all the rest were killed?". Later on he inspected the wreckage of the two aircraft and found that, of the 4 cockpits, his alone had remained relatively intact. Although most of his friends attributed his being alive to luck, DeHaven felt that the intactness of the cockpit structure was the answer; thus was born the first concept of "crashworthiness."

It is worthy of note that although hundreds of combat pilots and observers died as a result of injuries sustained in crashing their disabled aircraft — not from gunfire wounds—there is no record of anyone, other than DeHaven, having given any consideration to the direct causes of injury and death in aircraft accidents from an engineering point of view.

After the war, during the '20s, DeHaven's interest in impact injuries was heightened by newspaper accounts of so-called "miraculous" escapes from death of persons who had attempted suicide or had fallen from great heights. As more and more information was gathered on these cases, the fact gradually emerged that free-falling humans often escaped serious injury or death if their velocity was partly checked by their striking some light, frangible structure, such as a strong clothesline. Also effective for survival was their final impact in a supine or prone position on an object sufficiently "soft" to bring them to a stop through a relatively long distance, such as would occur when falling into a fire net, onto a light sheet metal duct or soft rubble.

DeHaven's insistence that these miraculous escapes were not miracles at all but were due to known physical laws relating to force per unit area (psi) received little consideration. In some quarters, DeHaven was even looked upon as a "crackpot." But he continued his studies on the phenomena associated with survival in free falls and, in addition, his attention was directed to injuries sustained in light plane accidents.

The results of these studies of small-plane accidents indicated that much could be learned about the direct causes of injury and, even more important, the reasons for survival under extremely severe conditions of crash force Thus was born a new concept, and a new profession – crash-injury research and crash-survival design engineering.

Impetus was given to crash-injury studies of small-plane accidents when DeHaven became associated with Dr Eugene D. DuBois at the Cornell University Medical College in New York in 1942; with the moral support of the Civil Aeronautics Administration and a small fund from the National Research Council, a statistical study of injuries and their causes in aircraft accidents was initiated.

During World War II, DeHaven, in cooperation with Dr. William Geohegan of the Cornell University Medical College, began experimenting with a device, later known as the inertia reel, which, used in combination with two vertical chest straps, would automatically restrain the torso and the head from flailing forward and striking rigid equipment in the cockpit. The development of this device had been due to the results of research studies which showed that most of the serious injuries sustained in aircraft accidents were confined to the head and the upper torso. Puncture wounds of the heart and the lungs, induced by penetration of fractured ribs, and *brain damage associated with and without skull fracture* were prominent results of accidents. Of particular interest was the fact that these types of injuries not only were sustained in completely disintegrated aircraft but also had occurred numerous times in cockpits and aircraft cabins which had sustained little damage. Studies of these latter type accidents, later to be termed "survivable" accidents, also proved that the so-called "dangerous" safety belt did *not* cut people in half. The studies showed further that, when decapitating wounds of the lower torso *had* occurred, they had been sustained in accidents in which the entire aircraft structure had been completely demolished; decapitation, it was shown, had been due to striking lethal objects such as jagged metal skin and fragmented, rigid wing and fuselage wreckage.

In the meantime, some farsighted individuals in the Bureau of Medicine and Surgery of the U. S Navy and in the Surgeon General's Office of the U. S. Air Force became interested in crash-injury research as a possible means of reducing the casualty

rate in the military services. Accordingly, a small contract through the Office of Naval Research was made with Cornell University Medical College to carry on DeHaven's studies of injuries in accidents. During the next 5 years, the Cornell group, consisting of a medical analyst, a stenographer and DeHaven, studied small-plane accident injuries.

To obtain information relating to the accidents concerning impact speeds, angles of impact, damage to the aircraft structures, and the medical histories of the occupants, special accident and injury forms were devised; since the Civil Aeronautics Administration was unable to provide anything more than token support (because the Civil Aeronautics Act did not provide for investigation into the causes of injuries—only the causes of the accidents), assistance of a few highly motivated state police and state aviation groups was obtained to conduct the investigations, complete the report forms and send them to Crash Injury Research for study.

Concurrent with these studies, the Cornell group developed a special punch card system and code for the coding of accident and injury data; a special injury scale, by which injuries could be assessed in relation to their seriousness from an engineering point of view, was also devised. This injury scale, which rates injury seriousness in relation to its danger to life under conditions of prompt and adequate medical care, proved to be of great value, providing much more information than the "loose" terms commonly used, such as "serious" and "fatal" (see end of chapter for Av-CIR Injury Code). The scale is divided into 10 parts, the last 4 of which are fatal. From an engineering point of view, with respect to design for survival, it is necessary to know the degree of death, i.e., whether 1, 2, 3 or more areas of fatal lesion had been sustained in combination with other nonfatal injuries.

During this phase of Cornell's crash-injury research studies, other groups and individuals became interested in impact injuries, particularly in relation to accidents involving military aircraft.

The work of Lieut. Col. John P. Stapp (USAF) in developing and operating a high-speed sled for studying high deceleration effects on human subjects is well known. Stapp's work, involving much personal courage, has added immensely not only in arousing world-wide interest in impact injuries but also in producing invaluable data for design engineers.

Two other military groups—the USAF's Directorate of Flight Safety at San Bernardino, Calif., and the U. S. Navy's Aero Medical Equipment Laboratory at the Naval Air Materiel Center in Philadelphia, Pa. began to investigate the human factors phase of aircraft accidents and produced information which showed that the human body could withstand, without serious injury, crash force of extremely high magnitudes, provided that crash-survival design concepts were utilized in the design of cockpits and restraining gear.

Other individuals and groups who began to sense the importance of studying the phenomena of crash-impacts and resulting injuries were William Stieglitz of Republic Aviation Corporation, A. M. Mayo, John R. Poppen and Stanley Lippert of Douglas Aircraft Company, Herman P. Roth and Charles F. Lombard of Protection, Inc., the USAF's Air Research and Development Command, the Aero Medical Laboratory, and the Cornell Aeronautical Laboratory, to name only a few.

By 1950, as a result of this progressive interest of an increasing number of engineers and safety people in the aviation field, a number of aircraft contained numerous features originally advocated by DeHaven's group. Forty G cockpits in fighter aircraft, lightweight, frangible (delethalized) instrument panels, control wheels and tilting seat-backs—the last to protect the heads of per-

sons in passenger cabins—were becoming standard equipment. Shoulder harness, which had proved itself of value during the latter part of World War II, also was being considered for light planes. Such terms and phrases as "crashworthiness," "survivable" and "nonsurvivable" accidents and the word "deceleration" became routine jargon of the industry.

Aside from military aircraft, increased crashworthiness also was being designed into some light planes by strengthening of the cockpit and the cabin structures, providing keel-like lower structures in the bellies of light twin-engine planes and moving the occupants further back in the airplane.

As the beneficial effects of crash-survival design in aircraft were becoming apparent, safety people in the automotive industry began to wonder whether this same design principle might not help to reduce the extent and the dangerousness of injuries sustained in automobile accidents.

Cornell's Crash Injury Research project turned part of its attention to the automobile accident-injury problem and initiated a research program in co-operation with the Indiana State Police. Following Cornell's lead, other groups and individuals soon turned their attention to this phase of highway safety. This program resulted in the growth of an almost nation-wide study of automobile accidents by many state police groups in co-operation with state medical associations, public health organizations and other safety groups.

The Traffic Institute of Northwestern University, the Institute of Transportation and Traffic Engineering at the University of California, The National Safety Council, Dr. Ross MacFarland of the Harvard School of Public Health, Motor Vehicle Research at Boston, Mass., the U. S. Army's Human Engineering Laboratory, and insurance companies were but a few who saw promise in designing for crash-survival.

Researchers in various parts of the country also began thinking about the use of safety belts and shoulder harnesses in automobiles; others were discussing the need for the crash testing of automobiles with dummies. Doctors in the medical research field, such as E. S. Gurdjian, J. E. Webster, H. R. Lissner and Jacob Kulowski began studying skull and skeletal fractures and brain injury. Scientists working under Edward Dye at the Cornell Aeronautical Laboratory began studying the kinematics of dummies subjected to decelerative forces, and the forces necessary to fracture simulated skulls when propelled against various structures.

At the same time, DeHaven and the author, who had joined the Cornell project in 1950, were endeavoring to interest the National Advisory Committee for Aeronautics in crash testing some small aircraft in order that decelerative loads imposed on the aircraft and their occupants under survivable crash conditions could be measured.

The NACA conducted these tests in 1953, and the results astounded safety people throughout the aviation industry. Loads of 30 G had been measured in these survivable test crashes of small, lightweight planes. Of even greater importance was the fact that crash-injury records showed that many persons had survived such crash loads with little or no serious injury. These data, plus that derived from many free-fall cases, also served to bolster DeHaven's contention that the human body could survive impact decelerations as high as 200 G without dangerous injury if adequate restraint and delethalization could be provided.

NACA's small-plane crashes also paved the way for later crash test programs of military fighter, bomber and civil transport aircraft. From these tests data have become available which will permit engineers to protect people more fully—by design—against the effects of impact loads imposed in survivable type accidents.

Early in 1950, Cornell's Aviation Crash Injury Research project centered its primary investigative attention on civil transport crashes. The resulting crash-survival studies on the Northeast Airlines' Convair 240 accident at La Guardia Airport, New York, and National Airlines' DC-6 crash at Elizabeth, N. J., interested safety engineering groups in the United States and abroad.

Both of these survivable crashes provided proof of the advantages of the crash-survival design features which had been built into these 2 transports and their seats—features which had resulted from crash-injury data obtained from small plane accidents.

Under the author's directorship, the Cornell Aviation Crash Injury Research project, while continuing to study crashes of civil transports, now co-operates with safety groups of the U. S. Navy, the Air Force and the Army, the Civil Aeronautics Board, the Civil Aeronautics Administration, and various other national and international groups, airlines and manufacturers. Each year sees a widening interest and understanding of the benefits that can be derived from crash-survival design.

An indication of the increasing awareness of the importance of this subject is shown by the fact that the Civil Aeronautics Administration revised its manual of procedure in 1955, wherein CAA safety agents now conduct crash-injury investigations on all survivable crashes involving aircraft in the nonairline category. These data are forwarded by the CAA to Cornell's Av-CIR for study.

Also, the National Advisory Committee for Aeronautics, for the first time in its history, organized a subcommittee devoted solely to flight safety. A large portion of this committee's work is involved with the problems of survival in relation to crash-impact.

The Airline Pilots Association—as a matter of policy—endorsed the need for, and the use of, shoulder harness for all airline cockpit crew members.

Seat manufacturers, both in the United States and abroad, are giving extensive consideration to providing greater safety through improved delethalization and better "tie-down" of the seats. Airframe manufacturers are endeavoring to integrate the seats with better hull and floor structures.

Consequently, the jet liners of tomorrow will contain a degree of crash safety unthought of 15 years ago.

In the automotive field, automobile manufacturers also are giving attention to designing for impact protection. Many items, such as safety belts, improved door latches, padded dashboards and visors, and energy-absorbing steering wheels, now offered in most cases as optional equipment, undoubtedly will become standard equipment on new model automobiles. This advancement in safety design in future automobiles will reduce drastically the high incidence of serious and fatal injury on our highways.

In effect, in the 14 years between 1942 and 1956 a new engineering field has been created, namely that of *crash-survival design engineering*. Undoubtedly, this subject will increase in importance as its value becomes more widely recognized. Thus, it is only a question of a short time before it will be taken up by universities and integrated into their design engineering courses. Once at this point, it can be expected that crash-survival design will increase rapidly in its application; perhaps it will show its effect in the design of objects in the factory, the office and the home which cause serious and fatal injury to people in accidental falls.

Only a brief glance at the number of serious and fatal injuries sustained by persons falling in showers and bathtubs points to the need of designing certain portions of our natural environment to protect the human body from injurious impact during "routine" accidents.

DEGREES OF INJURY*
TERMINOLOGY USED IN CRASH INJURY RESEARCH

†	‡	
1	1	*Trivial* or *None*
4	2	*Minor* "Minor" contusions, lacerations, abrasions in any area(s) of the body. Sprains, fractures, dislocations of fingers, toes, or nose. Dazed or slightly stunned Mild concussion evidenced by mild headache, with no loss of consciousness
15	3	*Moderate*—but not dangerous. "Moderate" contusions, lacerations, abrasions in any area(s) of the body. Sprains of the shoulders or principal articulations of the extremities Uncomplicated, simple or green-stick fractures of extremities and jaw. Concussion as evidenced by loss of consciousness not exceeding 5 minutes, without evidence of other intracranial injury.
28	4	*Severe*—but not dangerous. Survival normally assured. Extensive lacerations without dangerous hemorrhage. Compound or comminuted fractures, or simple fractures with displacements. Dislocations of arms, legs, shoulders or pelvisacral processes. Fractures of facial bones Fracture of transverse and/or spinous processes of the spine, without evidence of spinal cord damage Simple fractures of vertebral bodies of the dorsal and/or the lumbar spines, without evidence of spinal cord damage. Compression fractures of L-3-4-5. Skull fracture without evidence of concussion or other intracranial injury. Concussion as evidenced by loss of consciousness from 5 to 30 minutes, without evidence of other intracranial injury.
45	5	*Serious*—dangerous, but survival probable. Lacerations with dangerous hemorrhage. Simple fractures of vertebral bodies of the cervical spine, without evidence of spinal cord damage. Compression fractures of vertebral bodies of dorsal spine and/or of L-1 and L-2, without evidence of spinal cord damage. Crushing of extremities, or multiple fractures. Indication of moderate intrathoracic or intra-abdominal injury. Skull fracture with concussion as evidenced by loss of consciousness from 5 to 30 minutes. Concussion as evidenced by loss of consciousness from 30 minutes to 2 hours, without evidence of other intracranial injury.
66	6	*Critical*—dangerous, survival uncertain or doubtful. (Includes fatal terminations beyond 24 hours) Evidence of dangerous intrathoracic or intra-abdominal injury. Fractures or dislocations of vertebral bodies of cervical spine with evidence of cord damage Compression fractures of vertebral bodies of dorsal spine, and/or L-1, L-2, with evidence of *spinal cord damage. Skull fracture,* with concussion as evidenced by loss of consciousness, from 30 minutes to 2 hours. Concussion as evidenced by loss of consciousness beyond 2 hours. Evidence of critical intracranial injury.
91	7	*Fatal*—within 24 hours of accident. Fatal lesions in single region of the body, with or without other injuries to the 4th degree.
120	8	*Fatal*—within 24 hours of accident. Fatal lesions in single region of the body, with other injuries to 5th or 6th degree.
153	9	*Fatal* Fatal lesions in 2 regions of the body, with or without other injuries elsewhere.
190	10	*Fatal* Fatal lesions in 3 or more regions—up to demolition of body.

* Based on observations during first 48 hours after injury and previously normal life expectancy.
† Weighted value for degrees of total injury.
‡ Degrees of total injury.

4

Automobile-Barrier Impacts, Series II*

D. M. SEVERY AND J. H. MATHEWSON†

INTRODUCTION

Barrier crashes conducted in 1953 and 1954 furnished the first detailed information of the deceleration rates and patterns associated with ground vehicular crashes.‡ This type of experiment is easy to conduct relative to one involving two moving objects, since the impact parameters for the exacting purposes of research may be predetermined with reasonable accuracy. The rate of deceleration for the barrier crash generally exceeds that for other types of impacts at comparable velocities. Hence, the barrier impact provides a rigorous test having decelerations of the same direction and of comparable magnitudes and patterns as those which are encountered most frequently in collision-type accidents. Therefore, valuable basic data may be derived from experiments on restraining harnesses and similar safety devices under these conditions while simultaneously collecting much information on the collapse characteristics of the automobile. Because this type of impact eliminates the variables of (1) type of structure of opposing car, (2) mass of opposing car, (3) velocity of opposing car, (4) direction of impact and (5) point of impact, the barrier provides an excellent medium for testing the shock absorption effectiveness of force-moderating innovations and safety restraining-devices, all designed with the purpose of reducing the crash injury potential of the vehicular accident. These basic reasons indicate why the auto-barrier crash was selected as the first type of fully instrumented collision experiment to be undertaken by ITTE.§

EQUIPMENT AND FACILITIES

A barrier was constructed near the end of a 1,000-ft. dirt road (Fig. 1). This barrier, 8 ft. tall and 14 ft. wide, was made of large-

---

* Presented before the Society of Automotive Engineers, [illegible]

† [illegible], igi- neer [illegible]

‡ [illegible] in motor vehicle accidents is a nationally recognized problem it has been given increasing attention in recent years On what it believes to be a new approach, the Institute of Traffic and Transportation Engineering of the University of California, Los Angeles (hereinafter referred to as *ITTE*), began experimental studies of vehicle accidents in 1950. Initially, two-car head-on collisions were studied. Subsequently, the studies have included the phenomena measurable during impact between a single car and a fixed barrier. A series of such experiments have been performed and are continuing.

§ This kind of research, which originated at the University of California (ITTE), is an excellent example of the far-reaching effects that basic research usually has later on in the field of developmental progress. Certainly, these researches have given real meaning to the connections between crash episodes occurring in the external and the internal automotive environments. Already, automobile manufacturers are sponsoring similar types of experimental crashes in order to crashproof their cars more effectively. In this paper the reader is taken into the fascinating world of reality concerning automobile crashes by two masters of this sort of experimentation. Thus are we learning the why, the how and the when of rider injuries during crash decelerations. [Ed.]

diameter electric-utility poles sunk to a depth of 8 ft. in the ground and backed by suitable cross-members and braces to provide a rigid structure.

Cars of the same make, model and age were selected for these barrier collisions. Appropriate calibration markers were secured to the car (Fig. 2). Four flood-lights were mounted inside each car.*

A movie camera (modified G.S.A.P. 16-mm camera operated at 64 frames/sec.) was mounted on the rear shelf of the crash car and directed forward for the purpose of photographing the movements of the dummies during collision. Specially devised indicators were welded at 6-inch increments to the frame of the car from its forward end (left side) back to below the firewall. The target indicators of these devices were grouped behind the left-front wheel (Figs. 3 and 4). These devices provide data concerning the deceleration pattern for each 6-in. increment of the front section of the car frame. Accurate precrash and postcrash measurements were made of the frame, forward of the firewall, to provide permanent deformation data. The electrical leads from safety belt tensiometers, electric accelerometers, remote-control steering and braking devices, and other special equipment within the test car were joined to a cable which was

* The subsequent introduction of Tri-X film has made the use of interior lighting unnecessary.

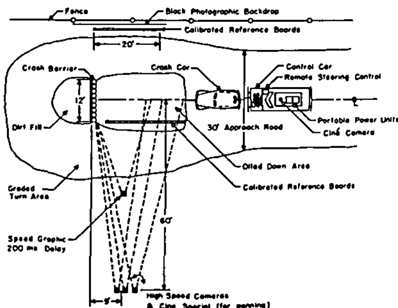


FIG. 1. The experimental site.



FIG. 2. Collision vehicle and instrument truck.

secured to an 8-ft. boom extending out from the rear of the car. This boom supported the cable during the crash, tending to prevent the cable from being thrown under the wheels of the car as the car rebounded from the barrier. This 100-ft. cable was connected to the push-truck which carried the recording oscillograph, the remote steering and breaking controls, the portable power units and accessory equipment.

EXPERIMENTAL PROCEDURE

The test car was brought up to the barrier to permit the photographic equipment to be

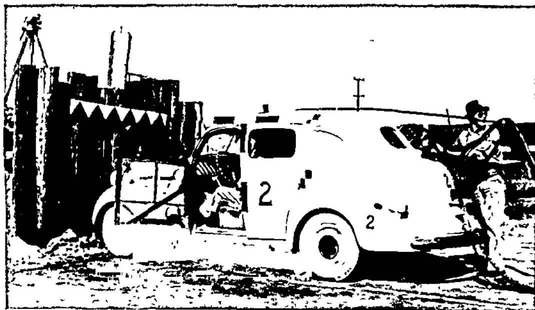


Fig. 3. Final adjustment of the multicable quick-release panel.



Fig. 4. Frame deceleration flag (lower left corner).

focused and directed at the vehicle. Following this, the vehicle was removed to a point approximately 1,000 ft. from the barrier. Then the instrument truck was used to push the test vehicle up to a steady speed of approximately 30 mph until a predetermined point in front of the barrier was reached. At this point the instrument truck was braked to a quick stop. The test vehicle coasted on into the barrier at an impact speed of 25 mph (Fig. 5). Throughout, the crash car was guided by remote control by an operator seated in front of the right section of the instrument truck windshield (Fig. 6). During the time the car took to coast this interval of 100 ft., the instrument truck was decelerated to a stop so that it was motionless, and the recording oscillograph was started before the car struck the barrier. Thus, the mobile recording station had become stationary in time to record the crash data without having the interference problems possible with mobile recording units.

INSTRUMENTATION

Photography

Two Eastman high-speed cameras (0-3,000 frames/sec.) were used as a precaution against possible loss of valuable data due to power failure, film processing accidents, etc.* Reference targets were painted on the principal points of interest of the car and on the dummies to provide a means, by high-speed photography, for recording data leading to the determination of velocity, acceleration, deformation and similar information. The G.S.A.P. camera mounted on the shelf above the rear seat provided an inside-the-car record of the dummies' reactions to impact. A Ciné Special movie camera was used for panning the car as it proceeded into the barrier. A second Ciné Special camera was mounted on the top of the instrument truck to provide a record of the collision in the direction of the motion of the colliding vehicle. Table 1 presents the photographic data related to these tests.

Camera-Oscillograph Synchronization

In order to provide a common time basis for relating the physical observations of the

* This precaution proved to be important because one of these two original films was damaged during the process of making a copy.

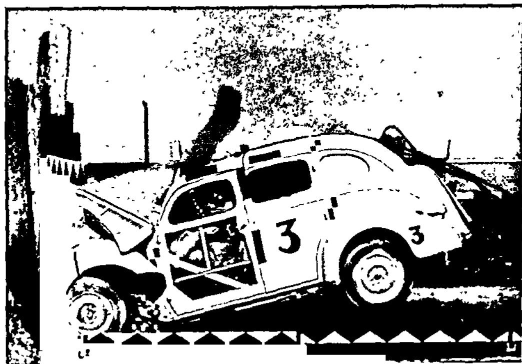


Fig. 5. The barrier collision at 29 mph.

crash recorded by the camera to the physical factors recorded by the oscillograph, a means of synchronizing these two variable speed-recording functions was introduced. This comprised feeding the signal from a magnetic pickup on the drive wheel of the camera to one channel of the oscillograph via a line from the camera to the instrument truck and dragged by the truck. The camera film speed was identified by a 60-cycle per second timing light marking on the edge of the film. The photographic paper speed of the Hathaway Recording Oscillograph was identified by the transverse line applied each 100th second.

The common time origin for these two recording systems was selected as the instant the front bumper of the car came into contact with the barrier. A probe was mounted so that it extended through the radiator flush with the front bumper. A slight axial movement of this probe actuated a switch which was connected to one channel of the oscil-



Fig. 6. Remote control driver for collision vehicle.

Table 1. Photographic Data Sheet

Camera	Position*	Type Film	f†	Shutter Speed (Frames/sec.)	Distance in Feet
Eastman High Speed—(ITTE), 16 mm.	60 ft. laterally 9 ft. longitudinally to impact point	Super XX (Eastman)	2.7	1,000	60
Eastman High Speed—(M I.S.), 16 mm.	Same	Super XX	2.7	1,000	60
G.S.A.P., 16 mm.	Back of car	Super X	5.6	64	8
Ciné I, 16 mm.	Top of Instr. Recording Truck	Super X	16	24	25
Ciné II, 16 mm.	Panning, near high-speed cameras	Super X	16	24	30
Speed Graphic	30 ft. laterally 9 ft. longitudinally to impact point	Super X	8	1/400	30

* Camera positions are shown diagrammatically in Figure 1.

† Weather was clear and sunny.

lograph. The broad end of the probe was clearly visible from the high-speed camera position so that the instant of contact also could be determined photographically.

Accelerometers

Eight electrical accelerometers were used, of which 6 were Hathaway units and 2 Statham. In addition, 5 Gross mechanical accelerometers were used. The characteristics of these devices are given in Table 2, while the placement and the axis orientation for these experiments are given in Table 3.

Experience has shown that aircraft, the automobile and vehicles such as the linear decelerator sled used by Stapp[5] developed vibrations within their structural components during collision decelerations which, although comprising seemingly negligible displacements, nevertheless may represent large accelerations. In such cases, the vibration of the structural member at its natural frequency may mask completely the total structure acceleration.

Table 4 presents the vibratory frequencies during the first 4 $\frac{1}{10}$ second intervals following the crash. The average of these values provides an estimate of the natural frequency of the structure on which the accelerometer unit was mounted. It appears that for the cars used in this crash, and for the structures instrumented, the natural frequency of these structures varied between 60 and 90 cps.

In many experimental studies, it is desirable to have detector units with as high a frequency response as is technically possible. However, in the problem of measuring decelerations of impacting structures, consideration must be given to the differentiation between oscillations and accelerations. Therefore, it is important to select a frequency response sufficiently high to reproduce adequately the impact decelerations without having the response so high that the unwanted vibrations (so-called "buzzes" of the structure) are included to confuse or even mask the basic data.

It has been estimated,[1] using a triangular pulse as a simplification of the decelerations pulse encountered during collisions, that with

Table 2. Characteristics of Accelerometers

Unit	Hathaway	Statham	Gross
Type	AMS—10A	F-10-120	Model C
Natural Frequency	230-260	120 cps	70 cps
Range	±15 G	±10 G	±50 G
Damping Coefficient	0.4 to 0.5	0.7	0.6 to 0.7
Damping Media	Oil	Oil	Oil
Functional Principle	Differential transformer, impulse fed to recording Oscillograph	Unbonded strain gage—impulse fed to recording Oscillograph	Jeweled Styli, cantilever mounted, on smoked drum rotated by spring action
Calibration Method	Dynamic and Static	Dynamic and Static	Dynamic and Static
Axis of Sensitivity	Perpendicular to base	Perpendicular to base	Parallel with base: bidirectional, records acceleration on 2 axes 90° apart

TABLE 3. DETECTOR UNIT DATA, CAR NO. 3

UNIT No.	CHANNEL No.	LOCATION	IDENTIFICATION	FIG. REF.	RANGE	DIRECTION
18	6	Head—SED (Dummy)	Hathaway Accelerometer	8	±15 G	Longitudinal
11	9	Chest—SED	"	"	"	"
19	12	Chest—ITTED	"	"	"	"
20	11	Head—SED	"	"	"	"
15	10	Top, Outside	"	4, 8	"	"
17	5	Door Post, Right Center	"	"	"	"
735	7	Chest—SED	Statham Accelerometer	"	±10 G	"
736	8	Door Post, Right Center	"	"	"	"
1	1	Chest Belt, SED, Left	ITTE Tensiometer	8, 12, 14	0-3,000 lbs	Parallel with belt load
2	2	Chest Belt, SED, Right	"	"	"	"
3	3	Lap Belt, ITTED, Left	"	"	"	"
4	4	Lap Belt, ITTED, Right	"	"	"	"
2	None (Mechanical)	Door Post, Top	Gross Accelerometer	—	±50 G	Long./Vert.
3	"	Door Post, Bottom	"	—	"	"
4	"	Frame, Forward	"	17	"	"
5	"	Frame, Aft	"	—	"	"
6	"	Roof	"	—	"	"

TABLE 4. NATURAL FREQUENCIES OF CAR STRUCTURE

	0.1	0.2	0.3	0 4	AVG.
CAR No 2					
Door Post (center) (Hathaway)	75	100	100	—	92
Door Post (center) (Statham)	65	60	60	65	62
Door Post (base) (Gross)	60	—	—	—	60
Door Post (top) (Gross)	60	—	—	—	60
Car Top (Hathaway)	65	80	70	55	67
CAR No. 3					
Door Post (Hathaway)	100	90	90	80	90
Door Post (Statham)	85	70	40	65	65
Door Post (top) (Gross)	70	—	—	—	70
Car Top (Hathaway)	75	65	60	60	65

an undamped natural frequency for an accelerometer of 50 cycles per second and a damping ratio of 0.7 or less, an accelerometer error of not more than 5 per cent results. Referring to Table 2, the natural frequency of the Hathaway accelerometers is 230 to 260 cps, and for the Statham, 120 cps. This relatively high frequency response appears to account for the 60 to 90 cps "hash" present in the deceleration curves produced from these units.

Hathaway Oscillograph

The signals from the various detector units mounted in the experimental car were fed to the 24-channel Hathaway Oscillograph Type S 8-C via its associated power supply units and its Strain Gage Control Unit, Type MRC-15 (Fig. 7). A sample of the data from this oscillograph is given in Figure 8.

Frame Deceleration Indicators

The mechanism of collapse of a structure during collision is of interest to the engineer. A knowledge of the order and the magnitude of deformation of each section of the structure provides a basis for evaluating the total structure in terms of its efficiency for attenuating the collision forces and therefore, the decelerations of the intact portions of the vehicle.

A system has been developed by ITTE for instrumenting each 6-inch section of the front of the car frame back to a point about opposite the rear edge of the front wheel. This system was necessary for the provision of accurate photographic instrumentation to a point on the structure obscured from view by the front wheel. This was accomplished by welding ¼-in. steel drill rods to the frame at these points in such a manner that they were nearly flat against the frame and pointed toward the rear of the car. A checkered target was painted onto the 3¼ x 3¼ x $\frac{1}{16}$ in. plate welded to the unsecured end of the rod (Fig. 4, lower left corner).

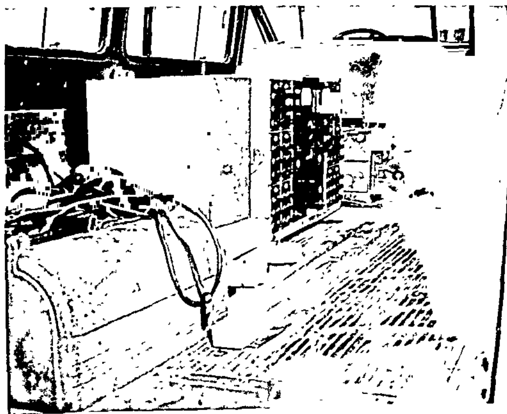


Fig nnel oscillogr

The movement of these targets as the car frame decelerates and deforms was photographed by high-speed cameras. The results obtained from this system of frame deceleration measurements is presented in a later section.

FRAME DEFORMATION INDICATORS

One method of obtaining useful engineering information from a collision is to measure specific car frame positions before and after impact in order to evaluate the location and the magnitude of collision deformations.

Positions on each side of the car frame were marked with metal screws at points approximately 1, 2, 3, 4, 5 and 7 feet back from the front edge of the bumper. The distances between these points were measured by projecting them onto a horizontal plane both before and following the collision. Details of this procedure are given in Refer-

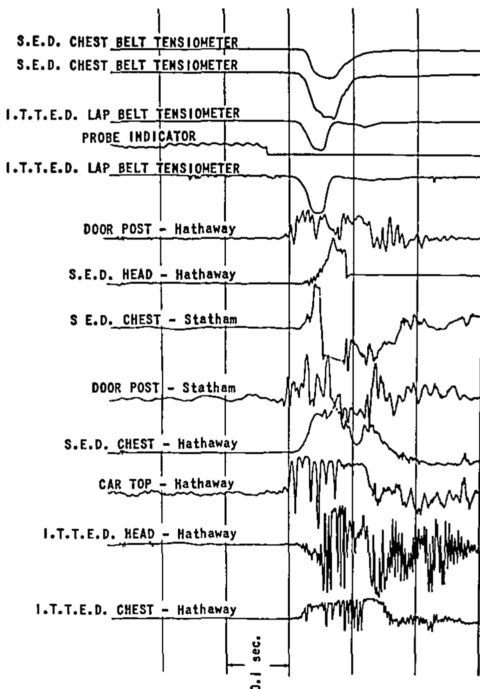


FIG. 8. Oscillograph record of collision with fixed barrier at 29 mph.



FIG. 9. Safety-belt tensiometer.

ence 3. The results of these analyses are presented under *Frame Deceleration and Deformation* in a later section.

TENSIOMETERS

Electrical accelerometers, high-speed photography, dummy-damage diagnosis and safety-belt tensiometers provided the instrumentation applied to the dummies. The belt tensiometers provided a force-time history of the loading to which the safety-belts were subjected. Four tensiometers were constructed for this purpose (Fig. 9). The belt is threaded between the 3 bolts connecting the 2 links. Application of tension to the belt bends the links sufficiently to permit the pair of SR-4 (Type A-5) strain gages bonded to each link to register the deformation. The tensiometers were calibrated statically, using a 60,000-pound Baldwin Universal Testing Machine. This calibration provided the basis for interpreting deflection of the recording oscillograph, connected to a belt tensiometer, in terms of pounds load. Tensiometers were used in pairs, one near each anchorage of the belt, in order to detect differential loadings to which the belt was subjected during collision. The records taken by these devices are presented in a later section.

PHYSIOLOGIC INSTRUMENTATION

Instrumented anthropometric dummies provide the most practical means of determining the injury potential to vehicle occupants under specific collision conditions. The data recorded during impact permits the determination of the force system applied to the motorist during collision and from this, the type, the nature and the severity of injuries likely to be encountered. An explanation of this system, together with the results obtained, is given in a later section.

The dummy* built by the Institute is referred to as ITTED, the abbreviation for Institute of Transportation and Traffic Engineering Dummy. The second dummy is called SED, after its manufacturer, Sierra Engineering (Company) Dummy. The Model 120 Anthropometric Dummy (SED) is described in detail by the manufacturer's brochure.[6]

In general, the more severe the collision conditions, the more closely the anthropometric dummy duplicates human responses, because the *small differences* between the dummy and the human relative to joint fixation, flesh compressibility and similar factors become negligible during *collisions* where the body may be loaded by 1 or 2 tons crash force.

EXPERIMENTAL FINDINGS

HUMAN BODY DYNAMICS WITH CHANGES IN BELT CONFIGURATIONS

The dynamics of the human body during a collision were determined for 5 conditions of restraint. Four restraints were evaluated by experimental collisions with a fixed barrier, using anthropometric dummies, while the fifth restraint was used by a human subject in a collision with a stationary car at 20 mph. The results of this latter experiment have been used as a basis for estimating the performance for that restraint in a collision with a fixed object at 25 mph.

*Anthropometric dummy. Sometimes referred to as anthropomorphic dummy. A mannequinlike structure having measurements, component weights, centers of gravities and similar dynamic parameters which correspond closely to those of the human body. The behavior of such a device during a crash or similar dynamic event corresponds closely to the kinematic behavior of the human body under similar conditions. [Ed.]

The effect of varying the belt configuration for a 25-mph collision with a fixed object is given in Figure 10. The performance of the belts used for the first 4 examples is discussed in connection with Figures 11, 12 and Table 5. The inferred injuries developed during the first 4 test conditions are presented in Table 6. The human subject using the combination restraint (lap and shoulder-loop belts) for a 20 mph-collision with a stationary car provided the basis for estimating the body dynamics given by the example at the bottom of Figure 10. Details of this test are described in connection with Reference 4.

Belt Tensiometer Results

Belt tensiometers provide a force-time history of the loading to which the safety belts were subjected. Tensiometers were used in pairs, one near each anchorage of the belt. (Figs. 11 and 12). For the shoulder-restraining device, 2 tensiometers were placed in series adjacent to each other in order to provide for evaluation of reproducibility of the tensiometer. Referring to Figure 11, the curves show the close reproducibility of information. An important characteristic of the shoulder-loop belt is the fact that the rate of onset and subse-

Table 5. Performance of 3 Belt Configurations

Restraint Configuration	Peak Force Lbs.	Belt Stress Duration in Seconds	Force Time Units	Relative Restraining Effectiveness*	Remarks
Chest	1,735	0.105	2,300	100	The chest belt dissipates a greater amount of body crash energy than the other (see below) 2 units without increasing the peak stress to the body. Physiologic considerations are discussed elsewhere.
Lap	1,725	0.057	1,500†	77	The lap belt is a somewhat less efficient restraining device for front-seat occupancy and for the front-end impact situation. It does not prevent the head and the upper torso from striking the forward surfaces of car interior. Energy is then absorbed from the most vital parts of the anatomy by injury-producing mechanisms.
Shoulder Loop	1,735	0 143	1,550	68	The shoulder belt dissipates the least amount of body crash energy. However, it maximizes (for a single restraint) the protection of the most vital parts of the anatomy, the head and the trunk.

* The area under the force-time curves (Figs. 10, 11 and 12) given in "units" by this table provides an index of the relative restraining effectiveness of each configuration. For ease of comparison, these values are divided by a constant to make the most efficient equal to 100. Consideration was given to the fact that lap- and chest-belt tensiometers were arranged in parallel so that their force-time values were additive, while the shoulder-belt tensiometers were arranged in series so that their values were averaged.

† This value must be corrected by the factor 200/170 to account for weight differences of dummies before determining the relative restraining effectiveness of the lap-belt.

TABLE 6. CRASH INJURY FINDINGS: AUTO BARRIER IMPACT
(Classes of Injuries: Minor, Moderate, Severe, Serious, Critical)

Car	Dummy	Forces on Safety Belt (lbs.)*	Deceleration of Dummy	Inferred Injuries†
				Head
No. 2 26 mph	ITTED Front Seat Passenger (wearing no belt) *Probable Fatality*	(No Belt)	Head: Calculated mean deceleration in excess of 200 G	CRITICAL Scattered abrasions; contusions around mouth, frontal skull and over occiput. Probable concussion. Fractured and lacerated nose. Combined injuries probably fatal.
	SED Driver (wearing shoulder loop belt) *Survived*	1,735	‡	MINOR Multiple abrasions. Minor contusions around nose, right eyebrow, around left earlobe, and chin. Other multiple small abrasions about head.
No. 3 Impact Vel. 29 mph	ITTED Front Seat Passenger (Wearing lap belt) *Probable Fatality*	1,725	‡	CRITICAL Multiple abrasions; contusions over entire forehead, nose, chin, frontal scalp (probable concussion—see Fig. 14) and occiput, neck and ears. Laceration right eyebrow, neck and head of model severed from model's body; inferred probable fracture of cervical spine.
	SED Driver (wearing chest belt) *Survived*	1,735	‡	MODERATE Contusion and laceration left forehead (eyebrow) contusion over posterior and midocciput, abrasion—right ear.

* Mean value of tensiometer peak readings for each belt taken from Figs. 11 and 12.
† Based on the examination and evaluations of Wendell Severy, M.D., Beverly Hills, Calif.
‡ The authors chose to delete these data because their accuracy could not be established. Structural buzzes confounded the basic data in the manner described in connection with Table 4. [Ed.]

TABLE 6 (*Continued*)

INFERRED INJURIES†		
TRUNK	ARMS	LEGS
CRITICAL Contusions and superficial abrasions over anterior midchest wall, probably a major crushing injury of chest because the instruments on chest were crushed; multiple contusion around pelvis.	MODERATE Abrasions right shoulder, lacerations left arm, upper and lower laterally; contusion right wrist. Laceration right hand, laterally and superiorly.	CRITICAL Left leg disarticulated at hip joint. Both knees disarticulated. Marked contusions with scattered abrasions right med. lateral thigh, left med. thigh and inguinal region medially; contusions below left knee and right knee.
MINOR Contusion midanterior chest wall (head snapped forward, driving chin into chest).	MINOR Few scattered abrasions both hands.	MODERATE Abrasion contusion left hip; deep laceration about 2 inches down to the bone in left upper medial leg just below knee joint—leg struck remote steering bracket (see Fig. 13).
CRITICAL Scattered abrasions; contusions upper anterior and right lateral chest wall; small contusion right shoulder; multiple contusions and abrasions lower anterior abdominal wall around pelvis; marked abnormal lordotic curve of thoracolumbar joint of model, inferred fractures of thoracolumbar spine.	SERIOUS Right arm dislocated from model at elbow joint; inferred probable fractures of right elbow joint. Right hand broken through midpalm, inferred multiple fractures of metacarpals, contusional right and left wrist. Fracture of left thumb	CRITICAL Both hips disarticulated from model; inferred bilateral fractures through neck of femur (bilateral). Severe contusion, laceration right lateral midthigh, inferred fracture midshaft right femur, multiple contusions left thigh anteriorly and posteriorly, left knee joint disarticulated from model; inferred fractures around right knee joint. Scattered contusions both ankles.
MINOR Contusion right upper anterior chest wall.	MODERATE Abrasion left hand (dorsum) and fingers. Fracture base of first phalange right third finger. Abrasion dorsum right hand.	MODERATE Abrasions, contusions and lacerations both knees medially just below knee joint. Abrasion-contusion and laceration right post. upper calf.

TABLE 7. COMPARISON OF DECELERATION CURVES OF FIGURE 15 FOR 3 COLLISIONS WITH A FIXED BARRIER

1	2	3	4	5	6
CAR No.	VELOCITY mph	AREA UNDER G-T CURVES SQUARE UNITS*	VELOCITY-AREA CORRELATION COLUMN 3÷2	RATE OF ONSET G/sec.	PEAK G
1	24.9	100	4.02	810	18.7
2	25.9	102	3.94	685	14.3
3	29.3	118	4.03	560	14.0

* Each square unit represents 1 G for 10 milliseconds.

quent reduction of the force is relatively gradual. *The belt restrains the body against significant forces over a comparatively long period of time.* These features of the curves suggest that the belt commenced restraining the body more quickly following the onset of collision so that large differential velocities could not develop between the car body and the human body.

Figure 12 presents tensiometer curves for

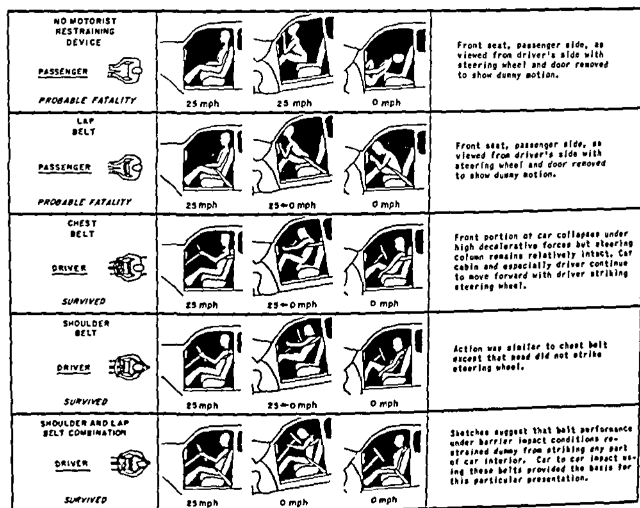


FIG. 10. Effect of varying safety-belt configuration for 25 mph collision with fixed barrier. All sketches on this page are reasonably faithful reproductions from high-speed motion pictures of actual collisions.

the lap belt and the chest belt. The tensiometers on the left side for both (front seat) dummies gave lower readings than those on the right side, indicating that frame failure on the left preceded the right side. This condition was unexpected, considering the fact that the collision was a direct (approximately perpendicular to the barrier) impact. The car was undergoing a very shallow turn to the right to correct its course toward the center of the barrier when it struck the barrier. Although the impact appeared to be perpendicular, the front side of the car frame may have been slightly leading the right side. Also, it is possible that irregularities in the barrier or the basic car frame structure may have accounted for this uneven application of forces.

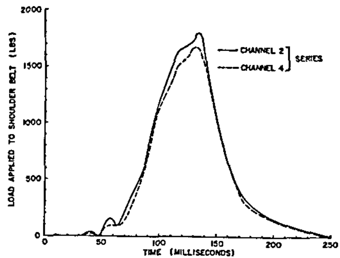


FIG. 11. Shoulder force—time history, S.E.D , car No. 2 (26 mph barrier impact).

The integral of the Figures 11 and 12 provides a basis for comparing the relative effectiveness of restraining configurations for the same collision conditions. Table 5 presents the results of this comparison.

The fact that peak forces for all 3 belt configurations were approximately the same must be regarded as coincidental, even though the cars were crashed under comparable conditions. This becomes evident when one considers the fact that this load is developed as a result of 2 contributions:

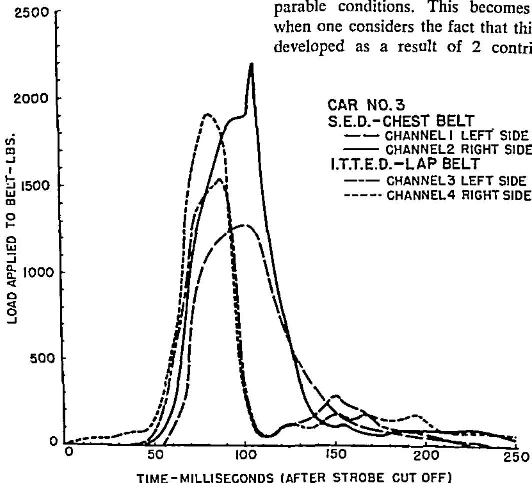


FIG. 12. Barrier impact at 29 mph.



Fig. 13. Anatomic pathologic diagnosis.



Fig. 14. Damaged head areas are immediately apparent because the bonded glass canvas covering turns white on being fractured.

the effective mass being restrained at the instant of maximum deceleration and the maximum deceleration to which this effective mass is subjected. Both of these factors will vary for the same impact event with variations in the restraining configuration so that the fact that the 3 configurations developed the same peak force under a condition involving a double variable must be regarded as coincidental rather than significant. This is further pointed out by an examination of the curves for each belt which shows slight variations in rates of onset and marked variations in durations of force.

Each belt configuration, per se, is capable of restraining effectively the body against the decelerative forces of this collision. However, particularly with respect to the front-seat installation, the effectiveness of each belt configuration is also dependent on the extent to which it prevents forceful striking of human body components against the car's interior during the collision event. To the extent that human body components dissipate their energy by striking the car interior surfaces, the belt tensiometers record correspondingly lower force values. Consequently, the excessive flailing of arms, the relatively smaller forces transmitted by the legs and the extremely destructive forces applied to the head and the chest during collision by the front-seat dummy secured by a seat belt account for the relatively small number of force-time units restrained by this belt configuration (Fig. 12).

If another column were added to Table 5 which included the potentialities of each belt configuration for reducing the severity of injury, then the shoulder belt and the chest belt would be upgraded considerably, relative to the lap belt, because of their performance of providing restraint to the vital portions of the anatomy (head and trunk). No attempt was made to include this factor in Table 5 because experiments were not conducted to determine the relative limitations of these configurations above which the belts themselves introduce injury.

Anatomic Pathologic Diagnosis

It is recognized that a medical authority cannot be expected to translate, without qualification, structural damage occurring to

an anthropometric dummy into injuries which the human counterpart would have been subjected. It is also impractical for an engineer to attempt to evaluate precisely the structural failures and other more subtle indications of abuse to which an anthropometric dummy is subjected during collision in terms of human injury data. Until more basic data is procured concerning the average strength and resistance to injury of the various components of the human body, the combined judgment of the medical and engineering researcher appears to be the most practical approach to applying this type of instrumentation to the experimentally crashed car (Fig. 13). These combined evaluations or interpretations of the physical consequences of dummy damage in terms of physiologic consequences of the inferred body injury are presented in Table 6 (see Chap. 7).

On the basis of the observations presented in Table 6, some conclusions are indicated. The reader is cautioned against regarding these findings as generalized or even specifically final in nature. A presentation of the inferred physiologic consequences of this particular type of collision has been made. While less exacting than most systems of instrumentation, it is believed that certain useful information is provided. However, it should be recognized that a change in any one of the many variables of collision may alter the injury pattern appreciably. Nevertheless, results of these tests do suggest the following:

1. For direct front-end impacts of the more serious type, the lap belt versus no belt for the front-seat passenger appears to offer little significant protection from injuries received by contact with the forward surfaces of the car (see Fig. 14 & Chap. 6). It should be pointed out, however, that the lap belt will prevent the body from being thrown out of the car or from being hurled about within the car in those rather prevalent collisions where spin-type forces are present. The serious consequences of such accidents suggest that the use of lap belts is extremely important. If it can be made acceptable to the motoring public, an upper torso restraining device should be developed.

2. The horizontal chest belt configuration continues to give experimental results which indicate satisfactory protection from serious impact injuries. This belt does permit the hips and the legs to shift forward in the seat so that the knees strike the instrument panel. However, this action is considered to be less serious than the action of the lap belt which allows the head and the chest to strike the forward surfaces of the car's interior. Before the use of this belt can be recommended, it is essential that the physiologic consequences, if any, of the pressures applied to the chest be investigated. This can be accomplished by conducting tests on live ape subjects. The use of cadavers is not recommended because some of the potential injuries of concern involve trauma observable only in live animals.

3. The shoulder-loop belt has proved to be the most promising single-unit restraining device evaluated to date. This experimental unit was developed by ITTE for the purpose of motorist protection by the use of a single-unit restraining device. Certain anatomic components of the human body are essential to life, while others are convenient accessories to the function of living. Protecting the limbs, or at least the legs, from impact injuries by the use of a lap belt may compromise under some circumstances the chances of survival by exposing the vital parts of the anatomy—the head and the trunk—to destructive impact. The shoulder belt restrains the trunk and the head at the shoulder level, permitting abdominal and pelvic regions to shift forward. This exposure of the lower trunk to possible injuries by the steering wheel, while undesirable, appears to be a more satisfactory compromise than the exposure of the head and the upper trunk because of their relative importance to survival. With the feet against the floor, the legs are better able to restrain forward motion of the hips than the less strong arms, holding the weak rim of a

steering wheel, are able to restrain the head and the chest. As the body shifts forward, the knees strike the instrument panel, thus loading the femur in compression, which affords additional restraint to forward travel of the hips. The undesirable shift of the hips could be prevented if both lap and shoulder-loop belts were used, but this more complicated restraining system for the present appears to be out of the question for the motorist.*

As a conclusion to these remarks, it may be stated that:

1. In severe accidents the motorist is almost certain to receive critical injuries unless he is restrained to decelerate with the car, thereby shifting the abuses of impact from the human to the car structure.

2. The effectiveness of the lap belt as a single restraining device has been evaluated only for one accident type. This device prevents the motorist from being hurled destructively about within the car or ejected from the car. The driver remains behind the wheel where, by retaining or regaining control, he may prevent the occurrence of secondary impacts that actually may prove to be more injury-producing than the initial collision. These experimental studies point out that users of the lap-type belt should be cautioned against regarding this device as providing protection against injuries for all types of impacts. The observations of researchers at ITTE suggest that the lap belt will prevent fatal injuries from occurring for some types of impacts.

3. The experimental performance of the horizontal chest belt has been very good. It is nearly as simple to operate and provides a more positive restraint than the lap belt. It may not be as comfortable to wear, particularly for women motorists, and *the physiologic consequences of its force system as a result of collision still must be evaluated before its use can be recommended.*

4. Without the need for introducing a more complicated restraining system, the shoulder-loop belt shows promise of overcoming both possible injury-producing consequences of the chest belt and the need for upper torso restraint not provided by the lap-type belt. Nevertheless, this belt is still regarded as an experimental model, even though with the use of dummies it has been tested in 3 collisions with impact velocities ranging up to 50 mph, and with the use of human subjects, in 8 collisions to 25 mph.

* However, it should be pointed out that this combination of restraining devices was chosen as the means for protecting human subjects in a recent series of automobile collision tests conducted at ITTE and as reported in Reference 4.

Head Movement During Impact

Analysis of high-speed motion picture film revealed that the head of both the human and the dummy passed through a 1.5 cycle oscillation. For both the human and the dummy restrained at the upper torso, the head appeared to be forced as far forward as the neck would flex during the first phase of the impact. After the car had decelerated nearly to zero velocity, the restitutional forces of the neck exceeded the now small deceleration forces of the head, applied through the neck, and the head was thrown back into an acute dorsiflexion position. Next, with the velocity of the car now at zero, the restitutional forces which had developed in the collapsed portion of the car accelerated it from the barrier in the reversed direction. This acceleration from zero velocity to 5 feet per second in combination with the forces of elasticity of the neck, in the case of the 25-mph collision, forced the head all the way forward again, though less abruptly than in the case of the initial forward movement. The resilient properties of the neck brought about the return of the head to an approximately erect position. Thus, the possibilities of whiplash injuries of the neck appeared to exist for the motorist who is restrained against a more violent trauma.†

† Note shearing forces at midbrain mentioned in Chap. 7. [Ed.]

While making the car considerably safer for rear-end collision exposure, this problem could be alleviated simultaneously if the seat backs were extended to provide head support in the vicinity of the seat where the head needs support against acute dorsiflexion.

ENGINEERING

The Problems of Establishing Experimental Controls for Collision Research. These problems are shown in the differences in the graphs of Figure 15. The significant characteristics of these curves are given in Table 7.

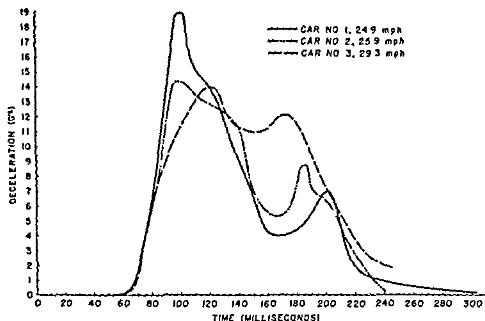


FIG. 15. Deceleration characteristics of the same portion of car body during collision with fixed barrier.

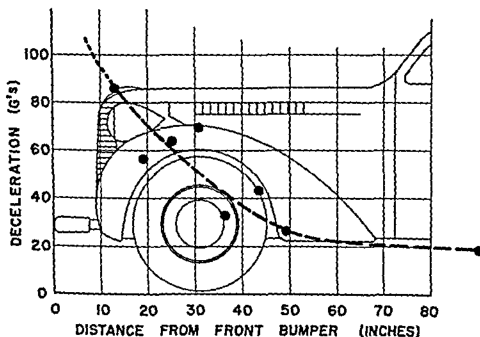


FIG. 16. Moderation of collision forces by the front-end structure at 25 mph impact with fixed object.

steering wheel, are able to restrain the head and the chest. As the body shifts forward, the knees strike the instrument panel, thus loading the femur in compression, which affords additional restraint to forward travel of the hips. The undesirable shift of the hips could be prevented if both lap and shoulder-loop belts were used, but this more complicated restraining system for the present appears to be out of the question for the motorist.*

As a conclusion to these remarks, it may be stated that:

1. In severe accidents the motorist is almost certain to receive critical injuries unless he is restrained to decelerate with the car, thereby shifting the abuses of impact from the human to the car structure.

2. The effectiveness of the lap belt as a single restraining device has been evaluated only for one accident type. This device prevents the motorist from being hurled destructively about within the car or ejected from the car. The driver remains behind the wheel where, by retaining or regaining control, he may prevent the occurrence of secondary impacts that actually may prove to be more injury-producing than the initial collision. These experimental studies point out that users of the lap-type belt should be cautioned against regarding this device as providing protection against injuries for all types of impacts. The observations of researchers at ITTE suggest that the lap belt will prevent fatal injuries from occurring for some types of impacts.

3. The experimental performance of the horizontal chest belt has been very good. It is nearly as simple to operate and provides a more positive restraint than the lap belt. It may not be as comfortable to wear, particularly for women motorists, and *the physiologic consequences of its force system as a result of collision still must be evaluated before its use can be recommended.*

4. Without the need for introducing a more complicated restraining system, the shoulder-loop belt shows promise of overcoming both possible injury-producing consequences of the chest belt and the need for upper torso restraint not provided by the lap-type belt. Nevertheless, this belt is still regarded as an experimental model, even though with the use of dummies it has been tested in 3 collisions with impact velocities ranging up to 50 mph, and with the use of human subjects, in 8 collisions to 25 mph.

Head Movement During Impact

Analysis of high-speed motion picture film revealed that the head of both the human and the dummy passed through a 1.5 cycle oscillation. For both the human and the dummy restrained at the upper torso, the head appeared to be forced as far forward as the neck would flex during the first phase of the impact. After the car had decelerated nearly to zero velocity, the restitutional forces of the neck exceeded the now small deceleration forces of the head, applied through the neck, and the head was thrown back into an acute dorsiflexion position. Next, with the velocity of the car now at zero, the restitutional forces which had developed in the collapsed portion of the car accelerated it from the barrier in the reversed direction. This acceleration from zero velocity to 5 feet per second in combination with the forces of elasticity of the neck, in the case of the 25-mph collision, forced the head all the way forward again, though less abruptly than in the case of the initial forward movement. The resilient properties of the neck brought about the return of the head to an approximately erect position. Thus, the possibilities of whiplash injuries of the neck appeared to exist for the motorist who is restrained against a more violent trauma.†

* However, it should be pointed out that this combination of restraining devices was chosen as the means for protecting human subjects in a recent series of automobile collision tests conducted at ITTE and as reported in Reference 4.

† Note shearing forces at midbrain mentioned in Chap. 7. [Ed.]

While making the car considerably safer for rear-end collision exposure, this problem could be alleviated simultaneously if the seat backs were extended to provide head support in the vicinity of the seat where the head needs support against acute dorsiflexion.

ENGINEERING

The Problems of Establishing Experimental Controls for Collision Research. These problems are shown in the differences in the graphs of Figure 15. The significant characteristics of these curves are given in Table 7.

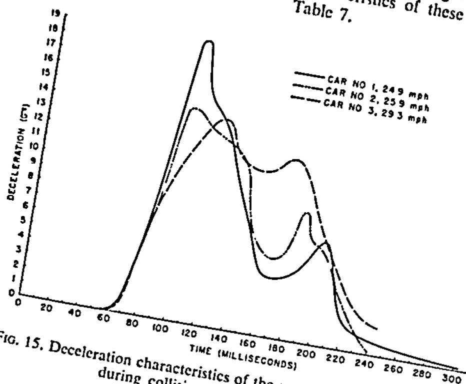


FIG. 15. Deceleration characteristics of the same portion of car body during collision with fixed barrier.

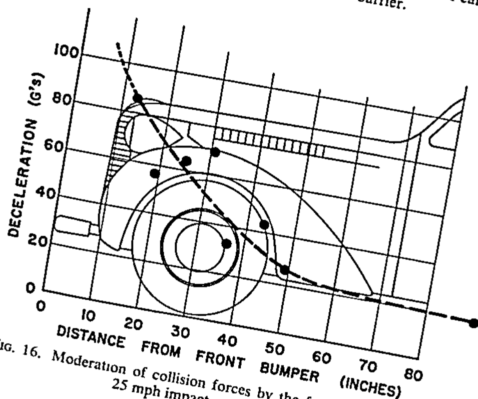


FIG. 16. Moderation of collision forces by the front-end structure at 25 mph impact with fixed object.

steering wheel, are able to restrain the head and the chest. As the body shifts forward, the knees strike the instrument panel, thus loading the femur in compression, which affords additional restraint to forward travel of the hips. The undesirable shift of the hips could be prevented if both lap and shoulder-loop belts were used, but this more complicated restraining system for the present appears to be out of the question for the motorist.*

As a conclusion to these remarks, it may be stated that:

1. In severe accidents the motorist is almost certain to receive critical injuries unless he is restrained to decelerate with the car, thereby shifting the abuses of impact from the human to the car structure.

2. The effectiveness of the lap belt as a single restraining device has been evaluated only for one accident type. This device prevents the motorist from being hurled destructively about within the car or ejected from the car. The driver remains behind the wheel where, by retaining or regaining control, he may prevent the occurrence of secondary impacts that actually may prove to be more injury-producing than the initial collision. These experimental studies point out that users of the lap-type belt should be cautioned against regarding this device as providing protection against injuries for all types of impacts. The observations of researchers at ITTE suggest that the lap belt will prevent fatal injuries from occurring for some types of impacts.

3. The experimental performance of the horizontal chest belt has been very good. It is nearly as simple to operate and provides a more positive restraint than the lap belt. It may not be as comfortable to wear, particularly for women motorists, and *the physiologic consequences of its force system as a result of collision still must be evaluated before its use can be recommended.*

4. Without the need for introducing a more complicated restraining system, the shoulder-loop belt shows promise of overcoming both possible injury-producing consequences of the chest belt and the need for upper torso restraint not provided by the lap-type belt. Nevertheless, this belt is still regarded as an experimental model, even though with the use of dummies it has been tested in 3 collisions with impact velocities ranging up to 50 mph, and with the use of human subjects, in 8 collisions to 25 mph.

Head Movement During Impact

Analysis of high-speed motion picture film revealed that the head of both the human and the dummy passed through a 1.5 cycle oscillation. For both the human and the dummy restrained at the upper torso, the head appeared to be forced as far forward as the neck would flex during the first phase of the impact. After the car had decelerated nearly to zero velocity, the restitutional forces of the neck exceeded the now small deceleration forces of the head, applied through the neck, and the head was thrown back into an acute dorsiflexion position. Next, with the velocity of the car now at zero, the restitutional forces which had developed in the collapsed portion of the car accelerated it from the barrier in the reversed direction. This acceleration from zero velocity to 5 feet per second in combination with the forces of elasticity of the neck, in the case of the 25-mph collision, forced the head all the way forward again, though less abruptly than in the case of the initial forward movement. The resilient properties of the neck brought about the return of the head to an approximately erect position. Thus, the possibilities of whiplash injuries of the neck appeared to exist for the motorist who is restrained against a more violent trauma.†

* However, it should be pointed out that this combination of restraining devices was chosen as the means for protecting human subjects in a recent series of automobile collision tests conducted at ITTE and as reported in Reference 4.

† Note shearing forces at midbrain mentioned in Chap. 7. [Ed.]

While making the car considerably safer for rear-end collision exposure, this problem could be alleviated simultaneously if the seat backs were extended to provide head support in the vicinity of the seat where the head needs support against acute dorsiflexion.

ENGINEERING

The Problems of Establishing Experimental Controls for Collision Research. These problems are shown in the differences in the graphs of Figure 15. The significant characteristics of these curves are given in Table 7.

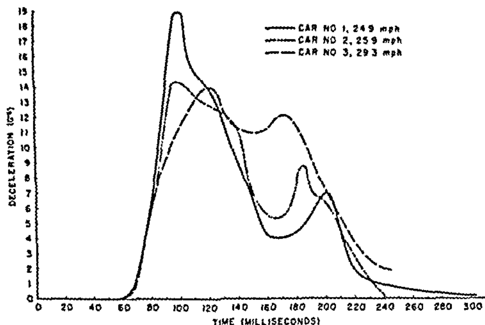


Fig. 15. Deceleration characteristics of the same portion of car body during collision with fixed barrier.

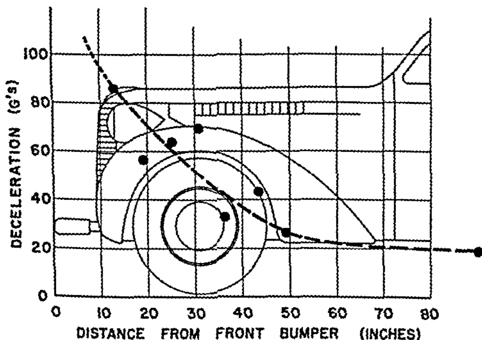


Fig. 16. Moderation of collision forces by the front-end structure at 25 mph impact with fixed object.

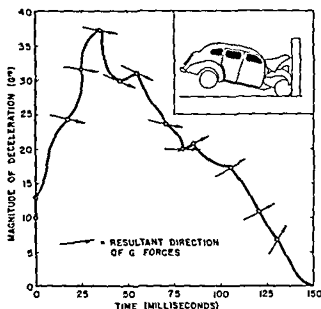


FIG. 17. Resultant direction and magnitude—car frame—left side, under firewall barrier collision.

COLLISION FORCE MODERATION BY CAR STRUCTURE

The average-priced American automobile has a structure which is rather effective in moderating the force systems applied during most collisions (Fig. 16). The somewhat irregular distribution of peak deceleration rates associated with the front portion of the frame is attributed, at least in part, to the presence of attachments such as the engine mounts. A homogeneous structure would be expected to fail with an orderly reduction of peak deceleration rates from the collapsed front-end to the undeformed portions of the car body.

RESULTANT CAR DECELERATIONS FOR DIRECT COLLISION OF AN AUTOMOBILE WITH A FIXED BARRIER

Collision deceleration is seldom unidirectional in that impacts usually do not involve contacts in which the direction of motion of their mass centers coincide. This means that rotational as well as translational accelerations generally develop during collision. Even direct impacts with fixed objects having large flat surfaces involve, in addition to longitudinal deceleration, vertical accelerations and decelerations.* The extent to which accelerations other than longitudinal are present is important, since they also have an influence on the motorist injury pattern. Figure 17 shows the resultant-direction and magnitude of the longitudinal and vertical decelerations for the car frame during direct impact with a fixed barrier. Initially, and during most of the collision, the decelerations are predominantly longitudinal but toward the end of the impact period a relatively strong vertical influence is evident. The sequence of strong longitudinal decelerations followed by smaller but increasing vertical accelerations may be fortuitous, since the motorist is not likely to be dislocated vertically before the high longitudinal decelerative forces are applied. This factor requires consideration when contemplating the use of certain restraining devices.*

Vertical accelerations have been observed for head-on and side impacts, but as yet no attempt has been made to determine their magnitudes and injury potential. The vertical acceleration values for the barrier collision apply only to this specific accident type. Vertical acceleration for a rear-end collision has been evaluated in Reference 4.

FRAME DECELERATION AND DEFORMATION

The timed-displacement of these checkered flags as the car frame decelerates and deforms is photographed by high-speed cameras (Fig. 4, area immediately to the rear of the front wheel). Since the metal flag is relatively weightless, and the rod was loaded essentially axial, the errors of measurement for this system may be expected to be low. However, a control for this instrumentation was introduced for the purpose of evaluating the error associated with deceleration flag measurements. A deceleration flag, hereafter referred to as the control flag (Target No. 10), was welded to the frame of the car about 1 foot in front of the door post and

* Head impacts with car top are not uncommon.

oriented so that the rod pointed back nearly flat against the frame. The rod was the same length as the longer units used in the front so that the error of measurement associated with this device would be representative of the error for the front devices. The checkered flag for this control unit also may be seen in Figures 4 and 6, against the frame under the rear door. The deceleration pattern observed for this control flag as contrasted with the reference (Target No. 9), painted onto the frame immediately in front of the welded end of the flag, is given in Figure 18.

The peak G of the control flag differs by 11 per cent from the peak G of the reference reading for the same point on the car structure. The areas under these curves representing the total force-time historys for that section of the car structure differ by less than 1 per cent. Thus the degree of accuracy of the family of deceleration curves obtained from 7 deceleration flags mounted at 6-inch intervals along the front third, left side of the frame has been established (Fig. 19). Of course, the specific values of peak deceleration for these curves will vary with the parameters of the collision such as, the speed, the type of vehicle, the object struck and similar factors.

In addition to the value of providing the design engineer with information relating to the problem of collision performance for the front third of the car, Figure 19 serves to indicate the kind of information on collision performance for front-end structures which may be derived from a relatively simple instrumentation technic. It is recognized that

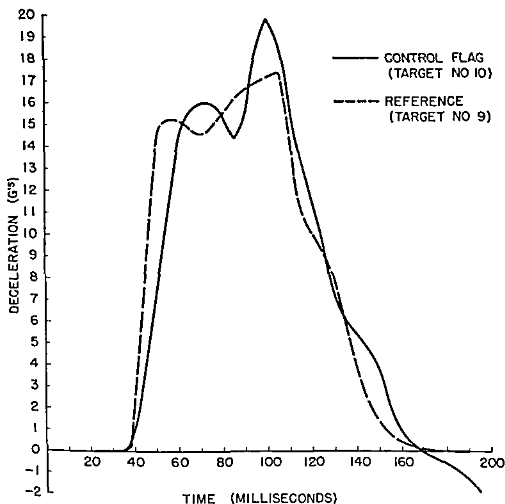


FIG. 18. Discrepancy between deceleration patterns for the control flag and the reference.

TARGET NO.	TARGET DISTANCE FROM FRONT BUMPER (INCHES)	PEAK G'S
1	13	85
2	19	56
3	25	64
4	31	68
5	37	33
6	43	43
7	49	26
9	91	17
10	91	20

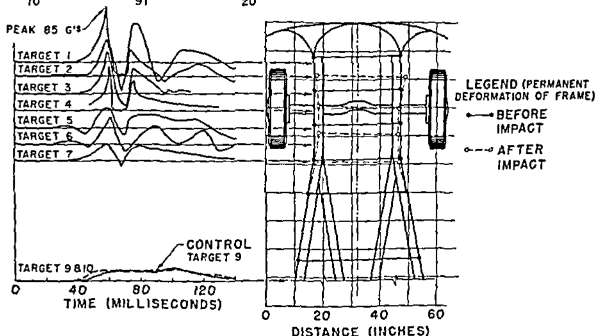


FIG. 19. Profile of deceleration during collision with a fixed object, 25 mph.

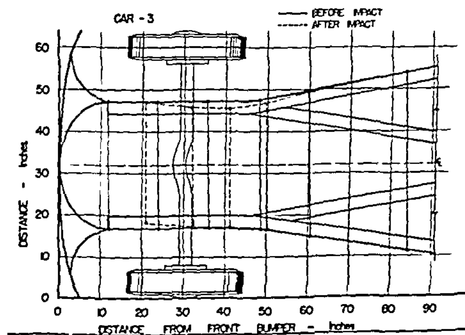


FIG. 20 ·rmanent deformation of frame for the 29 mph barrier impact.

following the development of this mechanical-photographic instrumentation technic at U.C.L.A., accelerometers capable of operating under severe impact conditions have been made available commercially. When the proper accessory equipment can be obtained, use of these instruments, although considerably more expensive, is recommended. The permanent deformations of the frame for one of the 25-mph barrier collisions are also provided by Figure 19 (right portion).

Three cars of the same make, model and year were crashed, independently into the same rigid barrier at approximately the same speeds. The permanent deformation of the frame for Car No. 1 (25 mph) was presented in Reference 3. Frame deformation for Car No. 2 (26 mph) is given in Figure 19, and for Car No. 3 (29 mph) in Figure 20. Cars 2 and 3 may differ slightly from Car 1 because in the case of the latter the 2 front seats were mounted on a pair of specially instrumented structural steel 4 in. x 8 ft. I-beams. The gross weight was 3,077 pounds for Car 1 and 3,380* and 3,240* pounds for Cars 2 and 3, respectively. A comparison of the deformation patterns for these 3 nearly identical vehicles crashed under essentially the same conditions is given in Figure 21. Reasonable correlation exists

* These two cars carried an additional dummy weighing 200 pounds which accounts for their marked weight difference.

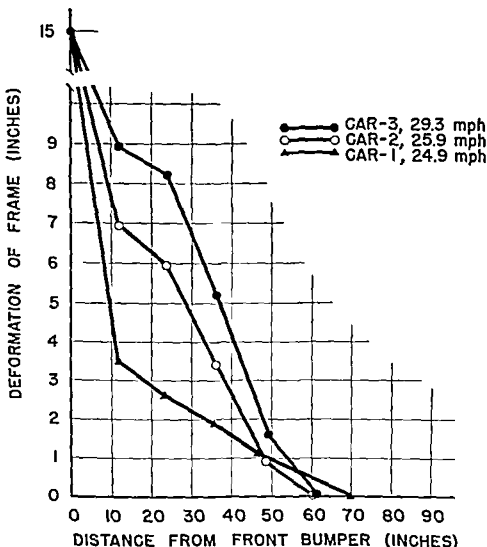


FIG. 21. Frame deformation patterns—barrier collisions.

TABLE 8. AUTOMOBILE IMPACT ANALYSIS

ITEM No.	DESCRIPTION	DERIVED FROM	CAR NO. 1	CAR NO. 2	CAR NO 3
1	Year and make	Data	1937 Plymouth	1937 Plymouth	1937 Plymouth
2	Body Type	Data	4-door Sedan	4-door Sedan	4-door Sedan
3	Measured weight with dummies	Data	3077	3,380*	3,240*
4	Velocity before impact	Data	24 9 mph (36.5 fps)	25.9 mph (38.0 fps)	29.3 mph (43.0 fps)
5	Velocity after impact	Data	—3 9 mph (—5 8 fps)	—3.1 mph (—4.6 fps)	—3.75 mph (—5.5 fps)
6	Deceleration rate (Peak for car top)	Data	605. fps^2 (18.7 G)	460. fps^2 (14.3 G)	449. fps^2 (14.0 G)
7	Duration of impact (Time for > 1 G)	Data	.170 sec.	.166 sec.	.19 sec. (approx.)
8	Amount of Crushing of car, elastic plus plastic	Data	2.3 ft.	2.4 ft.	3.6 ft.
9	Mass (w/g)	#3/g	95.6 slugs	105.0 slugs	100.6 slugs
10	Momentum before impact	#4x#9	3,490 lb.-sec.	3,990 lb.-sec.	4,330 lb.-sec.
11	Momentum after impact	#5x#9	—554 lb.-sec.	—483 lb.-sec.	—553 lb.-sec.
12	Total change in velocity	#4-#5	42.3 ft.-sec.	42.6 ft.-sec.	48.5 ft.-sec.
13	Kinetic energy before impact	½(#9) $(#4)^2$	63,700 ft.-sec.	75,800 ft -lb.	93,000 ft.-lb.
14	Kinetic energy after impact	½(#9) $(#5)^2$	1,610 ft.-lb.	1,110 ft.-lb.	1,520 ft -lb.
15	Coefficient of restitution	#5/#4	0.16	0.12	0.13
16	Change in momentum	#10-#11	4,040 lb.-sec.	4,470 lb.-sec.	4,880 lb.-sec.
17	Loss of K.E. during impact	#13-#14	62,100 ft.-lb.	74,700 ft.-lb.	91,500 ft.-lb.
18	Average force acting	#17/#7	23,800 lbs.	26,900 lbs.	25,700 lbs.
19	Average rate of energy dissipation	#18/#7	365,000 ft.-lb/sec.	450,000 ft.-lb/sec.	482,000 ft.-lb./sec
20	Crash horse power	#20(60)/33,000	664 hp	818 hp	876 hp

* These 2 cars each carried an additional dummy, weighing 200 pounds, which accounts for their marked weight difference over car No. 1.

TABLE 9. COEFFICIENTS OF RESTITUTION FOR AUTOMOBILE COLLISIONS

VEHICLE	IMPACT SITUATION	COEFFICIENT OF RESTITUTION
1937 Plymouth 4-door sedan	Collision at 25 mph with flat fixed barrier	0.16
1937 Plymouth 4-door sedan	Collision at 26 mph with flat fixed barrier	0 12
1937 Plymouth 4-door sedan	Collision at 29 mph with flat fixed barrier	0.13

between Cars 2 and 3. The removal of the 2 left side doors, and the alteration of the weight and the balance of Car 1 may account for its significantly different deformation pattern. Although only cars were selected which appeared not to have been

TABLE 10. COMPARISON OF 3 AUTOMOBILE BARRIER COLLISIONS

		TEST CAR 1	TEST CAR 2		TEST CAR 3	
Car: Make, Model, Year		Plymouth, 1937 4-door sedan	Same		Same	
Impact Velocity, MPH		24.9	25.9		29.3	
Weight of Car		3077	3380		3240	
Barrier*		Fixed Barrier	Same		Same	
CAR TOP	Peak G	18.7	14.3		14.0	
	Rate of onset, g/sec	810	685		560	
	Duration,† sec.	.240	.180		.240	
Coefficients of Restitution of Car		0.16	0.12		0.13	
ANTHROPOMETRIC DUMMY DATA	Dummy Used	ITTED	ITTED	SED	ITTED	SED
	Type Restraining Device	Horizontal Chest	Lap	Shoulder Loop	None	Horizontal Chest
	Peak Deceleration, G	24 5	‡	‡	‡	‡
	Rate of onset, g/sec	700	‡	‡	‡	‡
	Inferred Injury Classification	Minor, Judged survivable	Probable Fatality	Moderate judged survivable	Probable Fatality	Moderate judged survivable
	Loading applied to safety belt, lbs.	1800§	1725	1735	—	1735

* This barrier is illustrated and described in detail by Reference 3. (Also see Figs. 1, 3 and 6 of this study.)

† This is the duration for longitudinal decelerations in excess of 1 G.

‡ See footnote ‡ of Table 6.

§ This is an estimate based on the 170-lb. dummy being decelerated at rates up to 25 G with only the feet providing an added source of restraint.

involved in an accident, their history of use and abuse also may have a bearing on this divergence in correlation.

AUTOMOBILE IMPACT ANALYSIS

A comparison has been made of the physical factors of 3 collisions with a fixed barrier by practically identical cars crashed under essentially the same conditions. This data is presented in Table 8.

COEFFICIENTS OF RESTITUTION FOR AUTOMOBILE COLLISIONS

In the past, the coefficient of restitution for the crashing automobile was estimated as being nearly zero. Table 9 presents the coefficients from 3 U.C.L.A. automobile-barrier collision experiments. These values show that the coefficients may vary for the same accident type. Other U.C.L.A. experiments have shown that coefficients may be expected to vary for different accident types with a range from near complete plasticity to partially elastic reactions.

REFERENCES

1. Gerlough, D L.: Instrumentation for automobile crash injury research, Instrument Soc. America 1:29, 1954.
2. Severy, D . M., and Barbour, P.: Acceleration Accuracy: Analyses of High-Speed

Camera Film, Presented before the Society of Motion Picture and Television Engineers at Chicago, Ill., April 18, 1955.

3. Severy, D. M., and Mathewson, J. H.: Automobile-barrier impacts, Highway Research Board Bulletin 91, pp. 39-54, Washington, D. C., Nat. Acad. Sciences, January, 1954.
4. Severy, D. M., Mathewson, J. H., and Bechtol, C. O.: Controlled automobile rear-end collisions—an investigation of related engineering and medical phenomena (Series I: Montreal Conf. on Medical Aspects of Traffic Accidents, McGill University, Montreal, Canada, May 4, 1955), Canad. Services M. J. 11:727-759, 1955.
5. Stapp, John P.: Human Exposures to Linear Deceleration, Air Force Technical Report No. 5915, Part 2, Dayton, Ohio, Wright-Patterson Air Force Base, December, 1951.
6. Technical Report No. 120-901: The Design and Construction of Sierra Engineering Company Model 120 Anthropometric Dummy, Sierra Madre, Cal., Sierra Engineering Co., 1953.

5

Safety Glass: Past, Present and Future

WILBUR M. WHITE*

How many open automobiles did you see today? Or yesterday?

Today we ride in glass houses and we throw stones with comparative safety.

It is a result of a laboratory accident. A laboratory worker dropped his test tube of liquefied cellulose nitrate. The next day he found plastic hardened and adhering to the glass. A laminated glass had been born.

Fullicks, an Englishman, obtained patents in 1885, but there were no automobiles at that time to be glazed, so he thought of obtaining beautiful colors and combinations of colors and shadings for cathedral decorations. He used Canada Balsam to cement transparent celluloid between the layers of glass. Thus, laminated glass is 70 years old.

However, it was first presented for automobile safety use at the Spring Motor Show in England in 1906. Wood already had received the first patents for automobile safety use of laminated glass in 1905.

Much of the early laminated safety glass was used for goggles and gas mask lenses, as well as in airplanes and some in automobile glazing during World War I.

As for its automobile use in America, safety glass lay dormant until the late twenties when the rapidly developing closed car called for it. The first plastic used was cellulose nitrate with gelatin adhesive. Cellulose nitrate was strong, but stretched very little before tearing and proved to be unstable under sunlight. It turned brown with use, and its adjunct adhesives disintegrated.

In the early thirties, after much experiment and study, cellulose acetate was used and proved to be more stable, maintaining its clear color much longer. There still were problems with adhesives and stretchability.

A bulge break was desired with absorption of impacting energies.

Much is heard today of energy absorption in wreck impacts. The safety glass engineers were deep in the problem 25 years ago. They sat down and wrote a specification for a "made to order" plastic. It should be much stronger, with more stretch to absorb impacts without fracturing bones, especially skulls; and more stable chemically in sunlight; also, it should be its own adhesive.

It required time to attain this goal and almost endless research, not to mention costs. Above all, it took persistence and untold patience.

In the mid-thirties the chemists introduced the polyvinyls. However, so many workers had come close to the same thing that it required a pooling of developments and many patent compromises before it was practical to manufacture the material without resorting to litigation, endless debate and financial sacrifice; furthermore, the substance still was

* Executive Director of the Safety Glass Association, Inc, Hillsboro, Ohio.

EDITORIAL NOTE (J.K.). Mr. White's article on safety glass is instructive and provocative It places at the doctor's disposal for the first time a valid story in this regard. Mr White is one of the foremost authorities in this field; and, what is even more important, he is a great humanitarian who is trying to help solve some of the problems confronting those who are attempting to reduce automobile injuries under crash conditions and upsets

in the experimental stages. The new materials, of which polyvinyl butyral is most popular, have 300 per cent or more extensibility or stretch before tearing. The old plastics had about 13 per cent. For all practical purposes the polyvinyls have perfect chemical stability against sunlight. Few people realize what a destructive force the rays of the sun can be. Also, polyvinyl butyral has great strength.

It is a material well suited for automobile glazing. At no time since its development has any plastic appeared on the scene which has even come close to rivaling it. However, current studies to replace it have improved the polyvinyl butyral itself.

Among the difficult problems faced by the original makers of laminated glass was that of producing a glass only half as thick as the normal glass previously made. Now two sheets were to be used instead of one. This was especially trying in the case of plate glass. To grind and polish plate glass in very thin sheets resulted in great loss by breakage. Then too, the laminating technics were serious problems.

Understandably, bent windshields have posed many more problems. These have been met, and today their manufacture is almost routine.

Besides laminated safety glass, so-called "wireglass" is considered safe for certain uses. Also, heat-treated or tempered glass is acceptable for some purposes. Wire glass operates on the same general principle as laminated—that is, its safety function comes from holding the fragments in place when the glass is broken

Tempered or heat-treated glass acquires its safety qualities from the fact that the entire sheet tends to break into small fragments when its surface is ruptured. Its chief *safety factor* stems from *disintegration*, while the safety quality of laminated glass lies in its *resistance to disintegration*

Heat-treated glass is manufactured by first producing a glass pattern shaped and polished as for final use; next, it is placed in a furnace, brought almost to the melting point, then it is withdrawn and subjected to a sudden blast of cold air.

The result usually is a glass much stronger than ordinary annealed glass under cushioned blows or carefully loaded weights, but it is very fragile to concentrated blows or shocks.

It is the opinion of the writer that this material is too strong for use where impacts from human skulls are likely to occur. Skull fractures may result when the impacting velocity is over 15 miles per hour, experiments have shown with skulls impacting unyielding smooth surfaces.

An added factor in the deliberations of the American Standards Association's Committee Z-26, which placed limitations upon the use of tempered glass, was that of escape in event of wreck in water or fire. After long debate upon the question Mr. H. H. Allen, Safety Engineer for the Motor Carrier Division of the Interstate Commerce Commission, made a study of an extensive series of passenger-carrier accidents, after which he demanded limitation of its use. No person could break this material for escape purposes unless he were armed with a suitable tool.

Many suggestions have been advanced for use of plastic materials alone for motor vehicle glazing. Those advancing such proposals usually fail to consider the severe conditions that glass endures in motor vehicles, such as under windshield wipers. Grease, oils, sand, snow, ice, gravel, mud and almost every other class of grime must be removed by such wipers or by hand-rubbing on other glazings to maintain good clarity of vision—a "must" for safe automobile operations. A specially qualified group was appointed from the Safety Glass Committee of the American Standards Association to study comparative abrasion resistance of glasses and plastics. This subcommittee reported that *no plastic was as resistant within several*

hundred per cent as the worst glass when tested with the Taber Abraser, for example.

Grit or very hard sand is the worst offender under a windshield wiper. It can cut into glass in a few days if conditions are favorable. The toughest plastics maintain their surfaces only a few minutes under such conditions.

Adequate optical function of the windshield is of the highest importance in regard to driver visual acuity and other functions of "seeing." The question of what constitutes adequate optical function in this regard is a serious and complex one. Clarity of outline, minimum displacements of objects seen and "minimum jumping" of these objects are prime considerations. Of the three, no doubt the most important is "minimum jumping."

Clarity of outline adequate to ensure recognition of objects is necessary. Displacement of ½ inch in 25 feet is the limit allowed by the American Standard Code for windshield glass. "Jumping" objects or objects whose displacement is constantly changing or those which appear to jump is an intolerable fault in driving. It is caused by changing thickness of material through which the object is viewed or a change in the direction of "wedginess," such as the line of sight passing over increasing thickness, reaching the crest of increase and then going into thinner material. In such a case the viewed object seems to jump. The result is exaggerated nervous fatigue—a common cause of drowsiness and sleeping at the wheel.

Glass is the only known material that can maintain all these conditions and resist wind pressure to the point of keeping the same position in a glazing fixture such as a windshield frame.

A fact frequently missed by the novice in this field is that a plastic with a surface sufficiently hard to maintain its usefulness in windshields over a long period almost certainly would break with sharp cutting edges and cut just as raw glass cuts. Every test that the writer has ever seen in nearly a quarter of a century of intense observation in this field bears this out.

Now, what of the future? What is the outlook? Can the safety glass in use be improved? Are new materials developing to take its place? To say that the present product is perfect would be like saying the first few efforts at any job resulted in perfection. Such an outcome would exemplify a miracle of good luck.

The plastic used as interlayer in today's safety glass was developed after approximately 9 years of research by the whole automobile, glass and plastic industries. It was developed in answer to a well-designed formula for performance. It is practically tailor-made for its purpose.

Would different thicknesses of plastic or glass make a safer material? It is possible that it might, but tremendous sums have been and are being spent to find the answer to this question. Few weeks or days pass without tests of new thickness combinations. A perfect answer has not been found; or, if it has been found, this writer is not informed of the fact.

Men in the field of plastics maintain that they see nothing in plastic alone that promises a better solution of the problems than the existing combinations of glass and plastic. Of course, they investigate the possibilities of every new idea that occurs to them. Every big plastic company has hundreds of research specialists who know every angle of the problem.

To the genius who seeks to attain perfection, here are the specifications:

1. The glazing material must be dimensionally stable so that it can be permanently framed in another material so closely that wind and rain and storm are barred out no matter how wide the range of weather temperature or how great the velocity of winds from storm or driving speed.

2. It must maintain good visibility over a period of many years.

3. It must not discolor or turn opaque under a burning sun (which can tan even some glasses) within the lifetime of an automobile.

4. Any adhesive holding components together must last as long as the life of the vehicle.

5. It must keep its surface unmarred under the grinding of flintlike sands and gravels beneath the action and the pressure of a windshield wiper or cleaning.

6. It should yield under a blow from elbows, knees or skulls, but resist sufficiently, to absorb the forces of such blows and cushion them.

7. Sharp edges should be turned away from objects striking the material, and the edges should be dulled in the breaking action.

6

Kinematics of the Human Body Under Crash Conditions

EDWARD R. DYE*

When a vehicle is stopped suddenly, as in a crash, human bodies within the vehicle will also rapidly change their velocity to zero. The considerations of the complex and violent motions described by the various parts of the human body and of the forces associated with their deceleration are the subject of this discussion.

There are two general types. The first is that of unrestrained occupants. The second concerns the kinematics of the restrained body. The action of the two types is very different. In the first case (free body), the human continues forward at the onset of the crash until his forward flight is brought to a sudden violent stop on impact against the front of the car body. The free body is associated with a hammerlike impact blow where the duration of the arresting forces to the human is in the order of microseconds and the distance within which he stops is measured in fraction of inches.

The action is different when the human body is attached to his vehicle by an arresting harness of some kind. Now his body starts slowing down at the same time that the vehicle starts losing velocity. Thus, the stopping time is much longer, and the stopping distance is measured in feet instead of inches. Consequently, the forces on the human body are greatly reduced. Also, the motion path taken by the body or its parts is under control by the arresting gear.

For an engineering analysis of the motions, the human body can be considered as a complex linkage mechanism with all the link bars pivoted at the joints. All links, with the exception of the spinal column, can be considered as rigid bars from joint to joint. In our analysis, we considered the back as a flexible member. The stiffness of this particular bar was made proportional to the measured stiffness furnished by the tensed muscles of the backs of laboratory volunteers.

The motion of the compound figure is affected by both the weight and the moment of inertia of each of the connecting parts. As equations for the solution of kinematic motion of the human body are somewhat complex, we chose to use proportionally weighted, full-scale dummies as simulators

* Head of Safety Design Research Department, Cornell Aeronautical Laboratory, Inc., Buffalo, N. Y

EDITORIAL NOTE (J K) Edward R. Dye, an early disciple of Hugh DeHaven, states the basic geometric movements of the human body under forward crash (simulative) decelerative force conditions which he so ingeniously determined even in the days before actual barrier crash experiments had been developed. This truly pioneering laboratory [illegible] since instrumentation had not yet reached its present capabilities when he made his famed experiments with his "thin man," "thick man" and "half pint" dummies. Everyone engaged in crash-impact research is aware of his debt to Edward Dye and his [illegible]ified simulative experiments in regard to crash [illegible]lerative occupants.

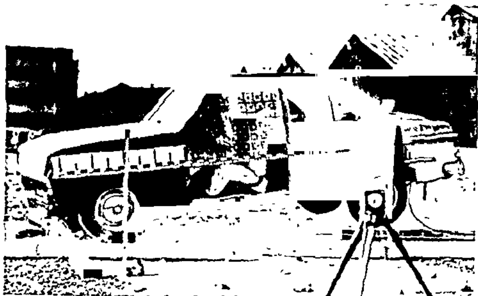


Fig. 22. Crash snubbing test vehicle. (Figs. 22-25: Cornell Aeronautical Laboratory)

to solve our problems quickly. The first studies were carried out on a full-scale proportionally weighted sheet-metal dummy distinguished from other laboratory dummies by the name "Thin Man." This dummy was designed to ride in a horizontal cockpit which was accelerated by elastic shock cords along a track on the floor of the laboratory. Time-spacial recordings were made on graph paper below the dummy through special instrumentation. The plotted data of some deceleration runs were made with loose joints to represent minimum muscular restraint. All were performed at the same 1.8 g deceleration rate (about 3 times that of frantic braking). This dummy and associated apparatus were made and used in 1948 primarily for the study of the effect of lap-type belts on the motions of the human and the relative effects of head blows with and without lap harness.

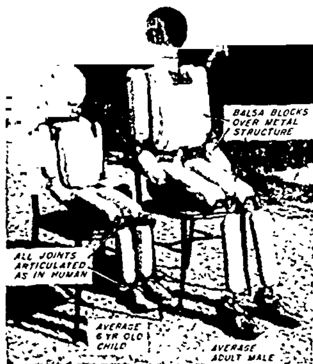


Fig. 23. Crash dummies (dynamically similar to man counterparts).

When no seat belt was used and the cockpit was subjected to a linear acceleration of 1.8 g, the head struck a point 27 inches forward of the head and 5 inches higher at a velocity of 16 ft./sec. and hit with an impact energy of 1,000 in. lbs. (600 in. lbs. is approximately skull fracture level against a hard flat surface).

When a seat belt was used at 30° from the vertical, and variously tightened to allow 4 and 2 inch movement respectively, the striking velocities were 15 and 17 ft./sec., respectively; but, the energies of the head blow were reduced to 380 in. lbs. and 240 in. lbs. respectively. When the arms are placed on a wheel or an airplane control stick and a seat belt is used, the energy of the head blow is still further reduced, probably because the mass of the arms is decelerated somewhat by forces through the

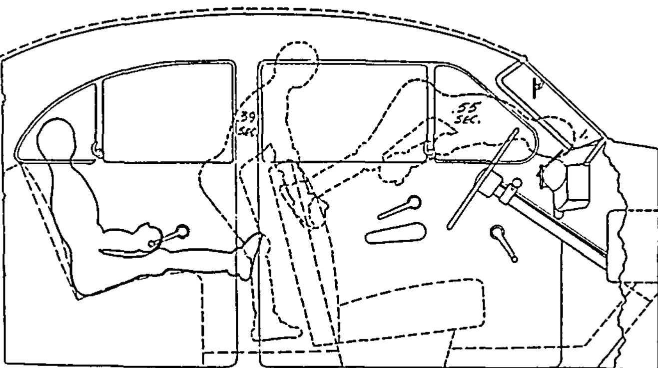


FIG. 24. Kinematics and hit locations for dummies. (Test No. 29. Car Velocity 27 fps. "G" stop 2.7. 6-year-old dummy right rear.)

hands. Also, the energies of the head blow were reduced to 40 in. lbs. and 160 in. lbs., *respectively*.

The "Thin Man" experiments were conducted with the cockpit being accelerated along a line without any rotation. This type of accident could occur when an airplane dives into the ground. The motions of the automobile body during an accident is more likely to include a rotational component.

In 1953, in a project sponsored by the Liberty Mutual Insurance Company of Boston, Mass., we made a study of the action of the human body in automobile crashes. Full-scale, proportionally weighted articulated dummies, one representing an adult and the other a 6-year-old child, were placed in a 1950 2-door sedan. The car was run down a 200 ft. track under its own power and then suddenly brought to a stop to simulate a crash. Through a transparent grid on the side of the car, we photographed the motions of the dummies as they moved through the car. All of these tests were simulating front-end collisions where the car entered the crash with the brakes set. Figure 22 shows the general action of the test vehicle during a crash snubbing test and Figure 23 the dummies used. The motion picture record was reduced frame by frame to give information concerning the flight path, the attitude of the body, the hit locations and the timing. Some of the test runs were of special interest.

The performance of a 6-year-old child in the right front seat when no seat belt was used showed almost pure translation until the lower part of the body was arrested when the feet and the knees came in contact with the car. Then there was a rotation about the hips until the head struck something—in this case, the windshield.

When a 6-year-old child in the right front seat was held in place with a seat belt, no blow was experienced.

Figure 24 shows the flight path of the child from the rear seat to contact with the instrument panel. It is interesting to note that the whole action took place in about ½ second even though the velocity of the crashing car was only 18 5 mph.

When the child was placed in the seat

face at 90° to the flight of the head produces the most energy of blow for a given crash velocity, about 78 per cent of the energy of the entire body. The kinetic energy of the body in ft. lbs. can be calculated by the equation

$$KE = \frac{WV^2}{2g}$$

Where W is the weight of the human body in pounds, V is the velocity of the car and the human at the onset of the crash expressed in feet per second, and g is the acceleration due to gravity in feet per second (approximately 32.2).

It is possible to use this sort of information in designing the interior of an automobile body to reduce the head injuries. For example, if all surfaces within the body of the car against which the head might strike could be shaped to offer a maximum angle of 60° to the head flight path, energy of head blows would be reduced to approximately 50 per cent.

The data on head impact locations under free body condition of the crash snubbing tests were reduced still further to get an envelope of possible head travel resulting from a front-end collision. We found that in front-end collisions the human head hit within a space 30° right or left in the horizontal plane and within a space 45° up and 30° down in the vertical plane. In the design of automobile bodies this zone should be kept free of objects having high injury potential.

Using combined information on the attitude of the body and the angle the barricade makes with the flight path of the head, calculations were made and data plotted for impact energy of head blows for both the front-seat and the rear-seat riders in a typical modern passenger car. These calculations were based on an assumed crash speed of 20 mph with the brakes fully applied (a condition typical in forward collisions) and the assumption that the car hit something solid enough to stop it in a distance less than the distance between the head of the person and the object in the car against which the head would strike. The weight of each occupant was taken as 200 lbs.

The approximate energy of the head blow at any location in the car can be determined for this set of crash conditions. This can be compared with the injury potential of any object within that zone to determine its hazard. Some data are already available on the head-injury potential of various objects and types of surfaces found in airplane and in automobile cockpits, and more information is being collected. As one would expect, there is a wide range of injury potential. For example, a 600 in. lb. blow to the head from a hard flat surface is required for head fracture, whereas only 60 in. lb. is required by a sharp 90° corner.

We are only beginning to scratch the surface in collecting useful engineering information on what happens to the human body in a car during a crash. We are limited by both manpower and money. Much is yet to be done, but the goal "to reduce the high injury and death rate on our highways" is well worth the effort.

7

Engineering Aspects of Fractures*

HERBERT R. LISSNER, M.S.†
and
F. GAYNOR EVANS, PH.D.‡

Skull fractures in automobile accidents occur less frequently than might be expected from a consideration of the energies and the velocities involved in crashes (Chap. 6). This does not mean that head injury is not a serious problem in automobile accidents. The relatively few skull fractures can be explained best after examining the mechanism of skull deformation and fracture production due to a blow.

All injuries are the result of the absorption of energy. Energy will be absorbed by the head whenever it comes in contact with an object that is moving at a different velocity.§ Three possible conditions may be described. First, the head may be at rest but free to move and be struck by a moving object. In this case the head will be accelerated. In the second condition the object may be at rest and be struck by the moving head. In this case the head will be decelerated (negative acceleration). The third possibility is when the moving head strikes or is struck by a moving object. In this case the head may be accelerated or decelerated, depending on whether the head or the object is moving faster and also on the relative masses and directions of each. For the 3 conditions described the injury to the head

* This research was supported (in part) by research grant A-377(C6) from the National Institutes of Health, USPHS.

Nowhere has the use of engineering principles of stress analysis been exemplified better than in the work and the writings of this school at Wayne University, as documented in the references at the end of this chapter. Their employment of instrumental technics has been done masterfully and has set the pace for others. This has been especially notable with the use of so-called "stresscoat" and electric strain gages There has been very little intrusion of mathematical manipulations to obscure the philosophic and clinical intent. In this chapter, two members of the Wayne team present and epitomize the results of many years of study based on original thinking and co-ordination of theoretical and clinical observations. Here, the connections between forces and fractures are summarized as they involve the skull, the femur, the lumbar spine and the pelvis. It is hoped that further studies will be extended to include other important parts of the bony skeleton The key to basic prophylaxis in crash injuries (to bone) should be found, ultimately, as a result of this sort of investigation. [Ed]

† Department of Engineering Mechanics, Wayne University, Detroit

‡ Department of Anatomy, Wayne University, Detroit.

§ The basic laws governing kinetics, or the relationships which exist between force, mass and acceleration (or deceleration), are embodied in the 3 Newtonian laws of motion which are learned separately but act together: (1) If the resultant force acting on a body is zero, that body will remain at rest or move with a constant velocity (in a straight line). (2) If the resultant force acting on a body is not zero, the body will be accelerated in the direction of the force, and the magnitude of the acceleration will be directly proportional to the force and inversely proportional to the mass of the body acted upon. (3) The force exerted by one body upon a second one is equal in magnitude and opposite in sense to the force exerted by the first body on the second one.

The first law illustrates that of inertia; the second is a quantitative statement (force equals mass × acceleration); and the third extends the second one to include aggregates of particles of bodies. [Ed]

will be identical, provided that the relative velocity between the head and the object is the same and the location of the blow on the head is also the same.

The absorption of energy* can produce mechanical damage to the head in the following fashion. (1) It can produce contusions or lacerations of the face or the scalp. (2) It can produce deformations and fractures of the bones of the face or the skull if the energy is sufficient. (3) The absorption of energy will produce pressure within the cranial cavity, this pressure being produced by deformation of the skull and the acceleration or the deceleration of the head.

Numerous studies have been made of the effect of blows to the head, evaluating these effects by means of engineering technics.[6, 12] These studies have included tests of both intact human cadaver heads and empty skulls at low and at fairly high velocities of impact. The tests were conducted to determine the extent and the character of skull deformation, the mechanism of fracture production and the energy required to fracture the skull. The time period during which the energy had to be absorbed to produce fracture was determined also.

Tests made at low velocities† of a magnitude encountered in ordinary automobile accidents, revealed the deformation pattern of the head due to a blow. In these tests an empty human skull was dropped on an unyielding flat surface of steel. The deformation pattern obtained may be described as follows: Directly under the point of impact‡ the skull tends to flatten out to conform to the shape of the surface against which the head is striking. In effect this produces an inbending under the blow due to the original curvature of the skull. After a certain amount of deformation has occurred the head begins to move away from the object which was struck as the inbended portion begins to rebound and assume its original shape. During the time when the point of impact was being flattened, the surrounding areas were undergoing a wave of deformation consisting of an alternate inbending and outbending taking place about the point of impact and at considerable distance from it. Thus, the point at which impact took place and certain radial areas about this point were bent inward, but at the same time adjacent radial regions were being bent outward.

In another series of tests the position of a single linear fracture produced by a blow at various regions of the head was determined. If the energy in the blow is sufficiently great, the region of the skull which is bent outward most severely will fracture. It is to be noted that this fracture begins at a considerable distance from the point at which the blow is struck. As the inbended portion under the blow returns to its normal position the fracture extends toward this region, as well as in the opposite direction, and may reach it and actually continue through it some distance.

To determine the location of the fracture due to a blow in any area of the head the skull was divided into 12 areas: 4 along the mid-line and 4 on each side. Blows were delivered to each of these areas in 100 ran-

* Energy of a body is the property that enables it to overcome resistance to motion, to do work, or to produce a physical effect.

† Mass, volume and time are scalar quantities; that is, having magnitude but no direction. On the other hand, acceleration and velocity are vector quantities, having both magnitude and direction. The latter distinguishes velocity from speed. Moreover, both acceleration and velocity are absolute values, while speed is related to some object in the environment. [Ed.]

‡ The principles of impulse and momentum clarify situations involving impacts and collisions between bodies. A linear impulse is obtained as the product of the force and the time interval of action. At any instant the linear momentum is the product of the mass and its velocity at that instant. A collision between 2 bodies which occurs in a brief interval of time and where relatively large reaction forces exist is called impact. Elastic impacts allow restitution of form of the impacting bodies. The longer the time interval, the less impulsive is the impact and the more crushing its effects. Also, the more acute the angles of impact become (carom), the less are the resultant forces (magnitudes). [Ed.]

domly selected adult skulls to determine where the initial single linear fracture would occur. Despite the large variation in the size and the shape of the skulls, a very marked uniformity of fracture position was found for the blows in each area. It must be remembered that the type of deformation and fracture obtained when the head receives a blow is a function of the velocity of the impact, the energy available and the shape of the object delivering the blow. In the tests described, the object delivering the blow was a flat steel plate. The results obtained from the tests indicated that a blow in the mid-line occipital region generally produced basal fractures. A posterior parietal blow in general produced fractures in the temporal region, and a midfrontal blow produced fractures above or through the orbits.

Tests at high velocities were made on empty skulls by dropping steel balls of various sizes onto the skulls through long distances. Various weights of steel balls were used to control the energy available when the skull was struck. As the higher velocities were reached the character of the deformation and fracture changed from that obtained with low-velocity blows. In the case of the higher velocity blows circular fractures occurred about the point of impact. The large deformations occurring at some distance from the point of impact found with low-velocity blows were no longer present. When high-velocity blows were struck with insufficent energy to produce fractures in both skull tables, it was found that the outer table was depressed, and the inner table remained intact. To confirm the single linear fracture obtained in low-velocity blows, steel balls also were dropped through shorter distances. These gave single linear fractures as in the case where the skull was dropped on flat surfaces

In order to determine the effect of the presence of the scalp and the skull contents, high-velocity blows on intact human heads were made with hammers. When the velocity of the blow was sufficiently high, localized fractures occurred under the head of the hammer. The presence of the scalp and its contents did not affect the results obtained.

The position of the single linear fracture due to low-velocity blows was confirmed by tests made on intact cadaver heads and also from the examination of clinical fractures. When the position of the blow was known in clinical cases the fracture position invariably fell in the region predicted by the tests on the empty skulls. The same sort of confirmation was obtained in the tests of intact cadaver heads. In addition, these tests were used to determine the amount of energy required to produce fracture and the time during which this energy was absorbed.

The energy required for production of single linear fractures with low-velocity blows was found to vary from 400 to 900 in. lbs. Although a slight variation occurred in energy requirements for fracture from one position to another within a single head, the variation was not great enough to be significant. Also it was found that the energy was absorbed in 0.0012 of a second. The first 0.0006 sec. was used in smashing the scalp, and the second 0.0006 sec. in deforming the bone. In these tests an SR-4 electric strain gage was cemented to the bone in the region where the fracture line was expected. As the fracture went through the gage it opened the circuit, and so the time period for fracture could be determined readily. The heads used in these tests were randomly selected adult cadaver heads weighing from 7 to 14 lbs. They were dropped on a heavy steel slab to ensure that all the energy would be absorbed by the head.

These tests also showed that the *scalp serves to absorb a tremendous amount of energy in comparison with that absorbed by the skull for fracture.* Empty skulls were fractured with 40 to 60 in. lbs. of energy in comparison with a minimum of 400 in. lbs. of energy required to fracture the skull when the head is intact. Another factor of considerable importance is that with only a

slight increase in energy absorbed, over that required to produce a single linear fracture, very much greater damage is produced. In other words, with a slight increase in energy beyond that required for a linear fracture complete destruction of the head results.

Examination of the specific conditions existing in automobile accidents reveals that frequently the victim is thrown from the car and strikes his head against the concrete pavement or some other nonyielding material. Under these circumstances the energy input into the head will be very large, and frequently fracture will occur. A free fall of only 6 ft. for a head weighing 10 lbs. gives an available energy of 720 in. lbs. If the head in this instance strikes an unyielding material or object, that 720 in. lbs. of energy will be absorbed by the head. In this case the head has a velocity of approximately 20 ft./sec. or about 13.5 miles an hour when it strikes. On the other hand, if the victim remains in the car during the accident and his head is brought to rest against some structure inside of the car, all of the available energy in the head, by virtue of its velocity, will not be absorbed by the head, but some portion of the energy will be absorbed by the structure into which it is thrown. The more deformable this structure, the less will be the energy that the head absorbs. As the head strikes, the structure or metal tends to wrap itself around the head, thus inhibiting the inbending of the skull at a particular point and so preventing fracture from taking place. The effect of the blow is no longer localized but is spread over a considerable area so that the skull is supported and fracture is prevented. For this reason skull fractures are not a common occurrence in automobile accidents in which the passengers remain within the car in survivable accidents.

However, bending of the structure to the shape of the head, which prevents the fracture, may prove to be quite dangerous. The prevention of fracture alone is not sufficient to prevent serious injury to the head. By decelerating the head at a rapid rate for a relatively long period of time, pressure is created in the cranial cavity. In turn, this pressure produces shear strains in the midbrain and the brain stem areas above the foramen magnum, resulting in concussion.[12] *There is no correlation between the severity of cerebral damage and the production of linear skull fracture.* Skull fracture may occur without damage to the brain; on the other hand, concussion and death may occur without skull fracture. Consequently, preventing the skull from fracturing does not ensure safety and protection for the brain. Single linear fracture and minimal concussion frequently occur with the same type

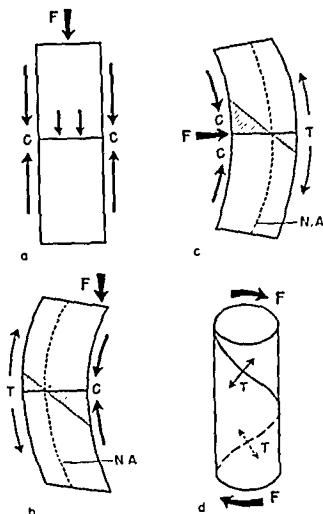


Fig. 26. Behavior of a column under different types of loading. C—compression stress; F—force or load; N.A.—neutral axis; T—tensile stress. (Evans: Am. Acad. Orthop. Surgeons Instructional Course Lectures 9:264-271, Ann Arbor, Edwards)

of blows. However, *if the energy input into the head is kept below 400 in. lbs., it is felt that neither severe concussion nor fracture will result.*

The most dangerous forces with respect to the mechanism of fractures of long bones are those which tend to bend or twist the bones. Such forces give rise to tensile strain within the bone which in turn causes linear fractures of the bone. The manner in which these forces produce tensile stresses and strains* in a long bone can be easily understood by comparing a long bone with a hollow column subjected to similar forces.[2]

When a column (Fig. 26 a) is loaded concentrically parallel with its long axis it is subjected to compressive stress and strain throughout the area of its cross section. However, when a force is applied to the column eccentrically, but still parallel with its long axis, it causes the column to bend as a result of which tensile stresses and strains are created on the convex aspect of the bent column while the opposite concave aspect is subjected to increased compressive stresses and strains (Fig. 26 b). Similar phenomena occur if the column is loaded like a beam perpendicular to its long axis (Fig. 26 c). In both of these types of loading the magnitude of the tensile and the compressive stresses and strains produced on the convex and the concave aspects, respectively, is greatest at the surface of the column and decreases internally until a neutral axis (N.A.) or plane is reached at which the magnitude of the stresses and strains is zero. If a column is subjected to a twisting or a torsion action (Fig. 26 d) tensile strain is created in the column and follows a spiral course around the column at approximately a 45° angle to its long axis.

That the gross biomechanical behavior of a long bone subjected to similar types of loading is a valid analogy to a column is evident from fractures experimentally produced in stresscoated femurs. "Stresscoat" is the trade name for a brittle lacquer which cracks in response to tensile strain in the underlying material.

Femoral fractures were produced experimentally by statically† loading the bones to failure in material—testing machines.[2,3,5] During these tests the load was applied slowly to the bone so that its behavior and the development of the tensile strain pattern could be observed accurately. In dynamic loading studies[2,5,14] in which the load is applied suddenly, the bone was loaded to failure by dropping a brass block upon it. The block was caught by hand on the rebound so that it struck the bone only once. In these tests the bone or part of it rested upon a heavy steel slab. The inch pounds of energy applied to the bone were determined by multiplying the weight of the brass block by the distance it was dropped.

An example of a transverse fracture of the neck produced by static vertical loading of a femur in the testing machine is seen in Figure 27. The fracture began on the superior aspect of the neck, the site where the Stresscoat cracks first appeared, and was

* The terms stress and strain refer to strengths of the tissues discussed, or mechanics of materials, and should not be confused with one another. Strain is the deformation which results from the application of external force and may manifest itself in linear direction, shortening as well as lengthening, volumetrically and morphologically. On the other hand, stress is the internal resistance to strain in the object or body which opposes the deforming action of the external forces. Stress and strain are related directly by a function of proportionality called the modulus of elasticity, which is a measure of the stiffness of a tissue or material.

Bodies have at least 3 forces acting upon them: *external (push or pull), inertial* and *gravitational*. Inertial forces act opposite to external or gravitational ones. The characteristics of a force which describe its external effect on a rigid body are its magnitude, direction of action and sense of the force along its line of action. [Ed.]

† Usually mechanics is subdivided into 3 parts: statics, kinematics and dynamics. Statics represents equilibrium between external forces and internal stresses. Kinematics is geometry of motion, without regard to forces or other causes. When the force ... the body ... oblem



FIG. 27. Transverse fracture of the femoral neck produced by a static load of 1,280 lbs. (Evans & Lissner: Anat. Rec. 100:159-190)

parallel with the direction of the cracks. As the load increased the fracture was seen to extend through the neck, final failure occurring after a maximum load of 1,280 lbs. had been applied. During this test the femur behaved like an eccentrically loaded column (Fig. 26 b). Also during the test the head and the neck of the bone were observed to be bent downward gradually by the slowly increasing load. This downward bending created tensile strain, as evidenced by the presence of Stresscoat cracks, on the superior aspect of the femoral neck in a direction parallel with the long axis of the region.

Further evidence of the behavior of the femur, like an eccentrically loaded column, was seen in the behavior of femurs subjected to vertical loading.[3] With a femur placed in a testing machine with just enough load to prevent it from falling (Fig. 28 a), a white thread extending vertically from the most medial point of the head of the bone to its intersection with the shaft was taut. However, when a load of 650 lbs. had been applied to the bone (Fig. 28 b) the thread became slack because of the bending of the shaft of the bone by the superimposed load. When the load was removed the femur returned to its previous condition, and the thread became taut again. This clearly indicates that the femur behaved as an elastic eccentrically loaded column.

Loading a femur perpendicular to its long

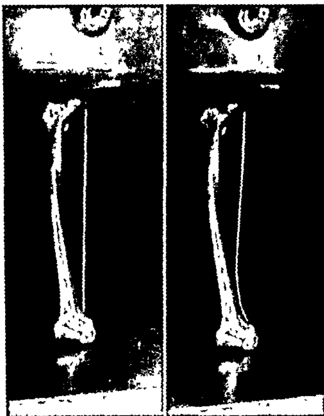


FIG. 28. Close-up view of femur in a testing machine. (a) Unloaded bone. (b) Same bone under a load of 650 lbs. (Evans & Lissner, Anat. Rec. 100:159-190)

axis also bends it with consequent tensile strain, as indicated by the appearance of the Stresscoat cracks, on the convex side of the shaft opposite that to which the load was applied. Under these conditions the femur is seen to behave like a similarly loaded beam (Fig. 26 c). When a sufficient load was applied to produce a fracture, the latter was observed to begin on the tensile side of the bone, that is, the convex side, opposite the side to which the load was applied. The fracture arose where the first Stresscoat cracks appeared and was parallel with the direction of the Stresscoat cracks, indicating, as in the case of the previously described fracture of the neck, that the fracture had arisen from failure of the bone because of the tensile stresses within it. Two fractures produced by loading the mid-point of the anterior aspect of the shaft are illustrated in Figure 29. The first cracks appeared on the convex aspect of the bone from the tensile

strain created in it as the bone was being bent by the load.

A variation of transverse loading of the bone perpendicular to its long axis was seen when the load was applied to the greater trochanter of the femur while the head of the bone was resting on a heavy steel slab. Under such conditions the bone was bent in a medial direction so that the inferior aspect of the neck and the adjacent medial aspect of the shaft were subjected to tensile strain, as indicated by the appearance of Stresscoat cracks in these areas of the bone. This is essentially what happens to a femur when one falls on the greater trochanter or when the distal end of the femur is abducted forcibly. For the latter reason the authors have called this "abduction" loading.

Various types of intertrochanteric fractures produced by static loading of the greater trochanter are indicated in Figure 30. Figure 30, 1, is an intertrochanteric fracture produced in a femur when it was supported by the head and the medial condyle. A different type of intertrochanteric fracture (Fig. 30, 2 and 3) was produced when the head of the bone was elevated 3 in. by a brass block. Crushing of the head occurred from above downward, but the fracture began at the lesser trochanter and gradually extended through the bone. The origin of the fracture line and the fact that it was parallel with the Stresscoat cracks indicated that it too arose from failure of the bone because of the tensile stresses within it. A third type of intertrochanteric fracture (Fig. 30, 4) was produced when the head of the bone was elevated 5⅜ in. by being rested upon a steel block. The fracture started inferiorly, in the region of the epiphyseal line, and spread horizontally to the posterior intertrochanteric line at a point midway between the greater and the lesser trochanter. Again, the origin of the fracture and the fact that it was parallel with the Stresscoat cracks was evidence that the fracture arose from failure of the bone because of the tensile stresses within it. Elevating the head of the bone reduced the effective bending forces acting upon the neck and the intertrochanteric region of the bone, while the direct compressive forces acting upon the shaft of the bone were increased.

During the abduction-loading studies little if any damage was noted at the site of loading of the greater trochanter. Comparison of these fractures with similar clinical fractures from the x-ray files of Detroit Receiving Hospital also revealed that commonly there is no apparent damage to the greater tro-

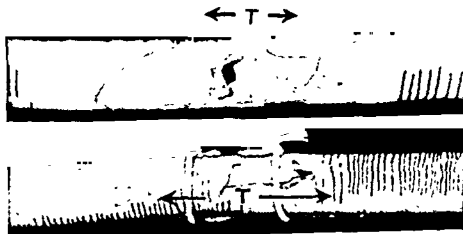


Fig. 29 Transverse fractures of the femoral shaft produced by a static load of 360 lbs. (*top*) and 390 lbs. (*bottom*). The load was applied to the center of the anterior aspect of the bone. T—tensile stress.

chanter. Structurally, the greater trochanter is a shell of compact bone filled with spongy bone. Although the magnitude of the force was sufficiently great to fracture the bone, the reason the greater trochanter rarely was crushed during a test is because of the excellent energy-absorbing capacity of the spongy bone filling the greater trochanter.

The tensile strain pattern produced by torsion loading of intact femurs (Fig. 31) is particularly interesting with respect to the mechanism of spiral fractures of long bones. The pattern as a whole spirals around the shaft of the bone at approximately a 45° angle to its long axis. The individual cracks composing the pattern also lie approximately at

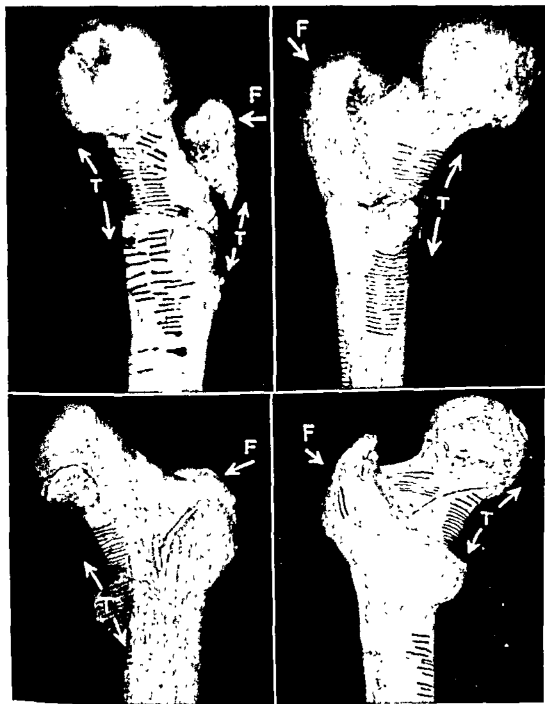


FIG. 30. Types of intertrochanteric fractures produced by static loading of the greater trochanter. F—force or load; T—tensile stress. (1) Fracture produced by a load of 1,290 lbs. (2) Fracture produced by a load of 1,365 lbs. (3) Anterior view of same bone. (4) Fracture produced by a load of 335 lbs.

a 45° angle to the long axis of the bone. The close resemblance between the distribution of this pattern and the behavior of a column under torsion loading (Fig. 26 d) is obvious. Spiral fractures of the femur produced by torsion loading (Figs. 31, 3, & 32) also take a spiral course around the shaft of the bone at approximately a 45° angle to its long axis.

The close correspondence in the direction of the fracture line with the Stresscoat pattern (Figs. 31 and 32) is again evidence that the spiral or torsion fracture of the shaft of long bones is simply another example of a linear fracture arising from failure of the bone because of the tensile stresses within it. Sometimes spiral fractures are stated to be the result of shearing stresses in the bone. However, if this were so, the fracture line would pass transversely across the shaft of the bone instead of spiraling around it.

The Stresscoat technic also has been used in studying pelvic deformations and fractures produced by static and dynamic loading.[4] In the static loading tests intact lumbar spines and pelves were loaded to failure in

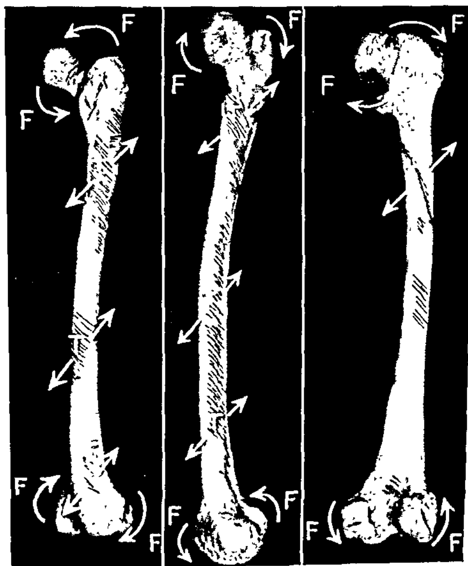


Fig. 31. Tensile strain patterns and fracture produced by static torsion loading. F—force; T—tensile stress. (1) Tensile strain pattern produced by 514 in. lbs. of torque. (2) Tensile strain pattern produced by 406 in. lbs. of torque. (3) Torsion fracture produced by 282 in. lbs. of torque

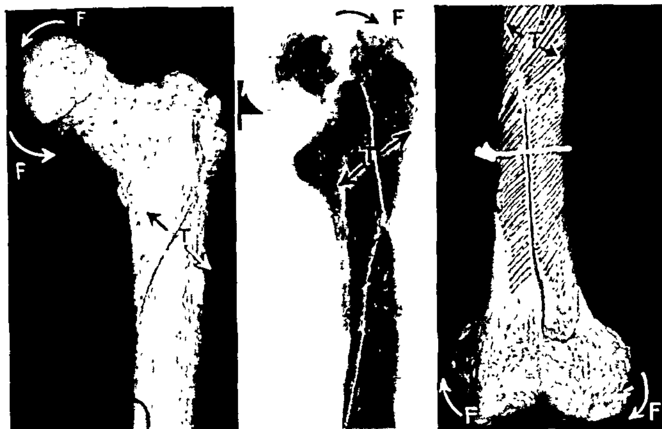


FIG. 32. Spiral fractures produced by static and dynamic torsion loading. F—force; T—tensile stress. (*Left*) Fracture produced by 448 in. lbs. of torque. (*Center*) Fracture produced by 498 in. lbs. of torque. (*Right*) Fracture produced by dynamic torsion loading. Magnitude of torque not determined.

a testing machine. In dynamic loading the intact pelvis was dropped upon a heavy steel block so that both ischial tuberosities struck the block simultaneously. The specimen was caught by hand on the rebound so that it struck the block only once. The weight of the specimen multiplied by the distance through which it was dropped gave the energy dynamically applied to it. This method of applying the load was chosen in an attempt to simulate the way the force is applied to the pelvis of a pilot during emergency escape from aircraft by means of an ejection seat.*

The biomechanical behavior of the pelvis responsible for the different types of tensile strain patterns (Fig 33) produced in dynamic loading tests is illustrated in Figure 34. Medial or lateral rotations of the anterior superior iliac spines gave rise to tensile strain parallel with the crest of the ilium in the iliac fossa or the lateral aspect of the iliac ala, depending on whether the rotation was in a lateral or a medial direction, respectively. If rotation of the iliac spine occurred in both directions the tensile strain pattern was found on both aspects of the iliac ala. Medial and lateral rotations of the ischial tuberosities gave rise to tensile strain in the lateral and the medial aspects, respectively, of the long axis of the ischiopubic rami. Rotation of the tuberosities in both directions resulted in tensile strain on both aspects of the ischiopubic rami. Lateral displacement of the acetabulum accompanied by posterior displacement of the symphysis pubis created tensile strain within the acetabulum as well as in the long axis of the iliopubic rami.

* The most severe fractures of the pelvis in crashes have been observed from forcible ejection; at the same time soft tissue injuries of the perineum and the genitalia as well as of the urinary bladder are also more likely to occur. [Ed]



FIG. 33. Tensile strain pattern produced by dynamic loading of the ischial tuberosities. (*Left*) Pattern produced by 208 in. lbs. of energy. (*Right*) Pattern produced by 85 in. lbs. of energy.

In order to determine the effect of the body weight, similar dynamic loading tests were performed on adult human cadavers from which only the lower limbs had been removed The deformations obtained closely resembled those of the previous tests with the isolated pelvis. An example of a fracture of the right ischium of a Stresscoated pelvis produced by 240 in. lbs. of energy is seen in Figure 35 a. Note the tensile strain within the acetabulum, the iliac fossae, the ischiopubic and the iliopubic rami as well as the lateral aspect of the ischium. The close parallelism in direction of the fracture with the Stresscoat cracks in the adjacent areas of the bone indicates that the fracture arose from failure of the bone because of the tensile stresses within it.

Figure 35 b shows multiple fractures produced in a pelvis by a statically applied load of 1,350 lbs. The general agreement in the location and the direction of the fracture

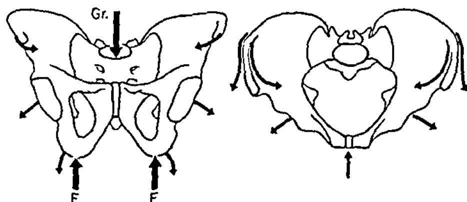


FIG. 34 Pelvic displacements under dynamic loading F—force; Gr - gravity Arrows indicate the direction of tensile stress. (Evans & Lissner Anat Rec 121:141-166)

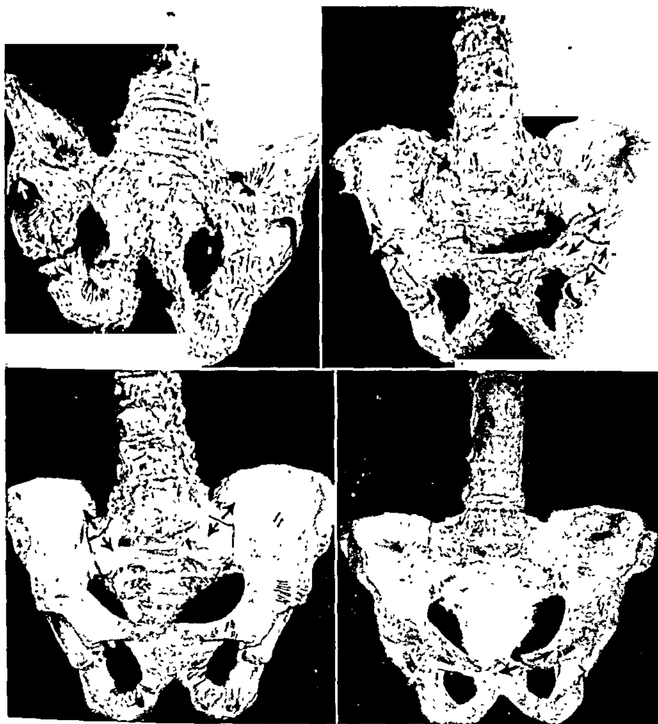


FIG. 35. Pelvic fractures produced by static and dynamic loading Arrows indicate direction of tensile stress. (a) Fracture produced by 240 in. lbs. of energy. (b) Multiple fractures produced by a static load of 1,350 lbs. (c) Fractures produced by a static load of 737 lbs. (d) A fracture produced by a static load of 112 lbs.

lines in this specimen with the Stresscoat cracks seen in the preceding specimen, as well as in other specimens tested under dynamic loading, shows that these fractures also arose from failure in the bone because of the tensile stresses within it. An example of a fracture of the right sacral ala produced by a statically applied load of 737.5 lbs. is illustrated in Figure 35 c. The Stresscoat cracks indicated in this specimen were produced by dynamic loading of the specimen and had no relation to this particular test. However, the parallelism of the fracture with Stresscoat cracks produced in corre-

sponding areas of other specimens (Fig. 33 b) suggests that the fracture also was a tensile failure. A fracture of the iliopubic ramus produced by a statically applied load of 112 lbs. is seen in Figure 35 d. This fracture is unquestionably a tensile failure because it extends transversely across the iliopubic ramus exactly parallel with Stresscoat cracks produced in this region by dynamic loading of the pelvis as seen in Figures 33 b, 35 a and c.

CONCLUSION

The conclusions reached from the Stresscoat studies of deformation and fractures of the skull, the femur and the pelvis is that the various linear types of fractures all arise from failure of the bone as a result of tensile stresses within it. These stresses are created by any forces which tend to bend the bone, and the fractures always are initiated on the convex or tensile side of the particular area of the bone involved. Therefore, any mechanism which reduces or lessens forces tending to bend the long bones or the pelvis will consequently reduce the tendency for linear fractures of these regions.

The experiments of Rauber,[15] Hulsen,[13] and Dempster and Liddicoat[1] all showed that the tensile strength of compact bone is considerably less than its compressive strength. Therefore, when a bone is subjected to both tension and compression stress it fails under tension. However, all fractures are not tensile failures. Compression fractures occur in the vertebral bodies and the calcaneum. All the various types of fractures experimentally produced could easily be duplicated by clinical fractures in the x-ray files of Detroit Receiving Hospital.

REFERENCES

1. Dempster, Wilfred T., and Liddicoat, R. T. Compact bone as a non-isotropic material, Am. J. Anat. 91:331-362, 1952.
2. Evans, F. Gaynor: Stress and strain in long bones of the lower extremity. Am. Acad. Orthop. Surgeons Instructional Course Lectures 9:264-271, 1952.
3. Evans, F. Gaynor, and Lissner, H. R.: "Stresscoat" deformation studies of the femur under static vertical loading. Anat. Rec. 100:159-190, 1948.
4. ———: Studies on pelvic deformations and fractures, Anat. Rec. 121:141-166, 1955.
5. Evans, F. Gaynor, Pedersen, H. E., and Lissner, H. R.: The role of tensile stress in the mechanism of femoral fractures, J. Bone & Joint Surg. 33-A:485-501, 1951.
6. Gurdjian, E. S., and Lissner, H. R.: Deformation of the skull in head injury. A study with the "stresscoat" technique, Surg. Gynec. & Obst. 81:679-687, 1945.
7. ———: Deformations of the skull in head injury studied by the "stresscoat" technique, quantitative determinations, Surg. Gynec. & Obst. 83:219-233, 1946.
8. Gurdjian, E. S., Lissner, H R., and Webster, J. E.: The mechanism of production of linear skull fracture, Surg Gynec. & Obst 85:195-210, 1947.
9. Gurdjian, E. S., Webster, J. E., and Lissner, H. R.: Studies on skull fracture with particular reference to engineering factors, Am. J. Surg. 78:736-742, 1949.
10. ———: The mechanism of skull fracture, J. Neurosurg. 7:106-114, 1950.
11. ———: The mechanism of skull fracture, Radiology 54:313-339, 1950.
12. ———: Observations on the mechanism of brain concussion, contusion and laceration, Surg. Gynec. & Obst. 101:680-690, 1955.
13. Hulsen, K. K.: Specific gravity, resilience and strength of bone, Bull. Biol. Lab. St. Petersburg 1:7-35, 1896 (in Russian).
14. Pedersen, H. E., Evans, F. Gaynor, and Lissner, H. R.: Deformation studies of the femur under various loadings and orientations, Anat. Rec. 103:159-186, 1949.
15. Rauber, A. A.: Elasticität und Festigkeit der Knochen, i-iv, 1-75, Leipzig, Wilhelm Engelmann, 1876.

8

Summary and Conclusions

JACOB KULOWSKI, M.D.

*The ever-increasing—even fantastic—pro*duction of American automobiles and other ground automotive vehicles, the consistently high level of actual gross crash injuries and deaths, despite lowered rates per 100,000,000 vehicular miles, has justified current considerations of motoring accidents in terms of epidemiology rather than as isolated mechanical events. From this standpoint, the injury (morbidity and/or pathology) is one thing, and its cause must be considered apart from causes of accidents themselves, which represent another thing entirely. Thus, a happy association has been developed between medical and engineering disciplines. Detailed studies have been and are currently underway which aim toward clarifying and co-ordinating human anatomy and human physiology with stress and strain analyses of vehicular structures and designs. Can humans and human parts be so studied under experimental conditions? Were the answer not in the affirmative, progress in modern high-speed transport would have ceased years ago.

Like all basic ideas, the statement that a crash injury is caused by bodily contact with a specific object or impact area in the internal and/or external automotive environments is stark in its simplicity. By the same token, its corollary, that the frequency and the severity of injuries can be reduced through *redesigning the injury—producing* environmental factors can hardly be doubted. Nevertheless, there are large areas, intervening between these two fundamental con*cepts, which beg for clarification before the* latter highly commendable aim in actual prophylaxis can be achieved practically. Here, the most elusive feature would seem to be the factor of the magnitude of forces which actually operate upon the body under crash and/or upset conditions. Perhaps the final answer to this question never will be found. Also, this final answer may not be necessary for our practical purposes of reduction or moderation of injuries during accidents. Certainly, enough has been said in the various chapters in this section of the symposium to indicate the validity of the crash-impact point of view. In other words, the basic ideas have been advanced. It remains for developmental progress to catch up with these experimental dicta. Changes in some aspects of current automotive engineering and design indicate that definite steps are being taken along these lines. It may very well be that eventually preventative medicine and engineering applied to the vehicular crash problem will eliminate these injuries from their present pre-eminence as a special category of mechanical injuries.[3]

However, before that wishful state has been reached, there is much that clinicians can do to help bring it about, in the areas of gathering and helping to interpret data on living morbidity and autopsy pathology. Fortunately, the first of these aims, the ac*cumulation of data, is not too difficult and* is being carried out by various individuals and groups. It remains only to co-ordinate these clinical data with their mechanical

*counterparts; i.e., a fusion of human and in*human elements into one basic design which reflects all of the physical factors involved in injury-producing accidents. In addition to the relationship which exists between bodily contacts and resultant force mechanisms of injuries which are received, the relative importance of the various bodily injuries must be recalled. Careful reflection upon these interrelationships between kinds of forces which operate to produce injury and the resultant lesions tends to focus the sharpest attention upon basic physiologic and pathologic engineering aspects which seem to govern the over-all picture, thus blurring out the purely clinical features of crash injuries. In other words, one can concentrate upon the primary injury and its cause or causes apart from all complicating sequellae because the latter hold no primary significance from this standpoint. Only on this broad spectrum basis and interpretation can available data be interpolated with engineering prophylactic endeavors.

It may be assumed safely that the kinematic behavior of the human body, as well as the human and the mechanical variables which are involved in crash decelerations and accelerations result in a multiple causation of injury production, although automotive design and magnitude of force are basic elements in it. The fact is that a combination of forces are acting upon vehicular occupants at all times—gravitational and inertial, in addition to the external ones derived from impacts between the body and the contact areas. Under crash conditions all of these forces tend to become exaggerated in general and more concentrated at particular points and areas of the body. Be that as it may, it is reasonable to group injuries generally in accordance with the forces which were probably responsible for them; i.e., applied forces (impulsive or crushing), inertial forces; and various combinations of these most likely to result from forcible ejection It is relatively simple to visualize injuries which are produced by *impacts and/or crushing forces.* Not infrequently, those caused by inertial ones are less distinctive, even to the careful clinical observer.

The most frequent inertial injuries currently being discussed involve so-called whiplash sprains of the neck and the low back. Until the recent work of the Wayne University school of observers, it was my personal opinion that these sprains had been receiving too much attention in the minds of the medicolegal fraternity. The fact that shearing stresses exerted in the region of the midbrain — subsequent to whiplashing — could cause cerebral concussion and/or cerebral damage must have a most sobering effect upon everybody who must deal with these injuries and their sequelae. Jolts, which are most frequently responsible for whiplash effects (chiefly rear-end collisions) can be moderated generally through changes in engineering design and materials which would tend to a more gradual (prolonged in time and distance) type of structural deformation under average loads of collision impacts. Thus, peaking would be minimized also, although this factor is probably not so prominent as was once thought to be the case.

More serious types of inertial force-injury relationships probably exist in respect to suspended organs of all kinds. A most spectacular inertial injury involves the partial or complete rupture of the aorta which is admittedly rare in riders of crash vehicles, but an excellent example of this sort of injury. The factor of safety suspension of the riders in ground vehicles should be discussed in some detail.

I am in complete agreement with those who advocate safety belts and other forms of safety barriers in order to afford greater protection to crash victims and increase their chances for survival. The words *protection* and *survival* have other important clinical implications which must be remembered as they relate to actual automobile occupants, not experimental subject or

anthropomorphic dummy. In the first instance, the forces are controlled in regard to time, direction and distance of deceleration. Moreover, the extremities are secured against flailing, etc. Finally, the subject is presumably a healthy specimen. There is no reason to doubt that safety barriers will give more protection and therefore increase the survival rate. But, it should be remembered that the chances for inertial types of injury (admittedly chiefly of a minor to moderate degree) are increased also when forces are being transmitted to riders through suspensory or retentive belts or harness. In other words, impacts and other applied forces are being substituted by inertial ones. It is a well-known fact that the areas of the body to which these latter forces are being transmitted are very tolerant ones. However, increasing protection and survival rate will result in some diagnostic and therapeutic problems which have not yet been given an opportunity to emerge clinically due to lack of time for, and the more widespread employment of, safety gear.

This statement is based upon autopsy findings in which obvious inertial injuries above and below the diaphragm were found in abundance but did not have any connection with the causes of death. In other words, had these people lived long enough, these tears and hemorrhages into omental and other tissues and structures would have produced symptoms to call for the most careful differential diagnostic thinking and treatment. A case in point is that of a state trooper who several months ago, rolled his car several times after a collision of moderate speed while he was wearing his safety belt. At that time he was hospitalized because of some abdominal discomfort and complaints. These vague but definite symptoms have persisted; the picture now being that of diffuse abdominal adhesions, presumably resulting from inertial injuries below the diaphragm. The clinician, a colleague of mine, is contemplating an exploratory abdominal operation for this man. It may well be that other reasons for the man's disability will then come to light. However, his complaints are all limited to the lower abdomen where the belt snubbed him violently.

Recently[1] the following specific crash-proofing suggestions have been made: (1) better roll-over frames or bars, at least double strength; (2) all-around safety belts as standard equipment; (3) safety door latches; (4) padded dashboard; (5) padding and restraining devices (for folding seats) for backs of seats, especially in second seat of station wagons, for children's protection; (6) eliminate rear seat window ledge, and consequently the placement here of dangerous articles; (7) smooth surfaces on the outside of cars (eliminate sharp dangerous projections to protect pedestrians); (8) better shock-absorbing (energy) steering controls; (9) hydraulic-type bumpers; (10) other safety features, including reflective tape on rear bumper and door edges, left outside mirror, back-up lights, better stop lights in general, electric signal lights, front window washers, photoelectric-eye light dimmer, drag-belt or static releasing chain, and a higher windshield and roof on some models for the tall driver. Many more will come to the manufacturers' attention as time goes on.

Oddly enough, the most obvious single injury potential in any automobile — the windshield—has received the least attention in recent years. Perhaps, the lacerative characteristics, as well as its contusive ones, of broken safety-glass have been dulled in the consciousness of current attitudes by the word "safety." This is one result of overworking a good word; even to the extent that the word or word sound actually takes over the significance of what it is supposed to mean. The intent and the action has been taken over by the sounded word. Actually, automobile manufacturers and safety-glass manufacturers are keenly aware of the shortcomings of so-called safety-glass and would like to improve it, and will do so when something better becomes available.

Another point worthy of special mention along lines of secondary safety has to do with safety automotive engineering maintenance. Up to the time that the automobile leaves the factory, its performance reliability is taken for granted by the buying public because this product has earned for itself that kind of acceptance from customers. The principle of *caveat emptor* no longer can be applied. The same cannot be said generally in respect to the reliability of garage service. It is mentioned here in the *sense that safety gear—for the present at* least—is for the most part optional equipment. Also, reliable performance is optional with drivers *but should be made mandatory.* Be that as it may, the servicing must be reliable safetywise. If it is not reliable, it is because the factory assembly is too complicated to allow adequate access to the vital parts of the engine, etc., and the fault is with the manufacturer who must remedy such a situation as soon as possible. In any case, manufacturers should see to it that their products are not impaired by poor servicing. Improvement in this area could do much to rejuvenate an ailing industry. However, it must be admitted that few accidents can be attributed definitely to mechanical failures. Nevertheless, the above admonitions fall into the over-all mosaic of the safety *pattern and cannot be ignored or neglected.* Also, strictly speaking, this is an area for accident prevention but is another example of how all facets of this problem tend to blur into one another in a complementary or supplementary fashion. This is true for the practical as well as the theoretical aspects of motoring accidents, injuries and safety

Progress in crash-impact engineering and design tends to show that human tolerances to crash forces—as they operate during accidents—are adequate enough for survivable crashes, but that current basic concepts of engineering are not taking full advantage of that human factor. Already, steps have been taken toward that end, and greater promises seem to be imminent. Thus, another link has been forged in the chain of approaches and methods of study and controlling motoring accidents in order to lower *the morbidity and the mortality. It has been* stated that driver proficiency—upon which all primary safety depends to the greatest single extent—is the result of psychosomatic-mechanical factors. Already, two of these elements have been dealt with; i.e., the somatic and the mechanical, albeit from the standpoints of the injury and injury cause alone.

Considerations of the psychosomatic-mechanical factors involved in accident prevention will bring us to Part III which will be published in the spring of 1957. That *will complete the ring reflecting the clinical* life history of motorists and motoring. It is a ring composed of several (3) separate rings like those in a puzzle; for flexibility and rearrangement whenever that is called for in the complex make-up of this modern activity of mankind.

REFERENCES

1. Gibbens, Murray E., and William V. Smith: The Doctor and the Automobile Accident, Paper presented at the annual meeting of the A.M.A., Chicago, June 11-15, 1956.
2. Kulowski, Jacob: The Clinical Aspects of Automobile Accidents and Injuries, exhibit at A.M.A. annual meeting at Chicago, June 11-15, 1956.
3. Stapp, John P.: personal communication.

Index